

SPENCER P. THORNTON, M.D.

OPHTHALMIC EPONYMS

*An Encyclopedia of Named Signs,
Syndromes, and Diseases
in Ophthalmology*



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EPONYMS

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Named Signs, Syndromes, and
Diseases in Ophthalmology

By

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Dr. Thornton is a graduate of Bowman Gray School of Medicine. He not only completed his residency in ophthalmology at Vanderbilt University, but also a residency in general surgery at the University of Alabama in Birmingham.

This work, beginning with Acosta's Syndrome and ending with Zinsser's Disease, and a selection of Surgical Eponyms, includes more than six hundred eponyms encountered in ophthalmology and related disciplines such as neuro-ophthalmology and pediatric ophthalmology. All synonyms are given for each eponym; and all are indexed. With each eponym, references to the literature are given. There are more than 2,000 cross references. To easily identify the name of the diagnosis of unusual cases seen in the office and clinic, Dr. Thornton has arranged the last section of his book as a diagnostic guide, beginning with abnormalities of the eyebrow, abnormalities of the lids, the canthus, eye position and ocular motility, and then considerations of the conjunctiva, the cornea, the iris, the lens and the structures of the fundus. Finally, the systemic components of the syndromes are topically indexed, rounding out this book as a prime diagnostic tool.

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Dedicated
to
Ginnie, Steven, David, Ray, and Beth
For many stolen hours

PREFACE

THIS book was compiled in response to a long felt need for a reference manual to the many ocular signs, diseases and syndromes eponymically designated in ophthalmic and general medical literature. It has been my purpose to present, in as concise a manner possible, those points of importance attached to the eponym by recurring references in the world literature over the past several decades.

The descriptions given are as used in the literature and not necessarily as originally described. Different writers place emphasis on differing aspects of a condition and hence one may find apparent conflicts in the description of single eponyms, and duplications in the case of multiple eponyms (see Batten-Mayou, Stock-Spielmeyer, Vogt-Spielmeyer). This can be appreciated further when one encounters eponymic references with multiple applications. For example, the term Sjögren's syndrome calls to mind most frequently the patient with the triad of keratoconjunctivitis sicca, dry mouth and arthritis. However, to the student of pediatric or hereditary ophthalmology, the term Sjögren's syndrome may call to mind the autosomal recessive combination of congenital zonular or polar cataract associated with oligophrenia, hereditary ataxia and other congenital anomalies.

The bibliography as listed is not intended to be always the original, the most recent, or the most descriptive. The references to the literature are given primarily for the interested reader who may want to see the term as it has been used by contributors to the literature. But where there have been differences in subsequent usage of an eponym, it was felt of interest to cite the original description in a number of instances.

Most eponyms bear the name of the man who presented the original description or who attracted the most attention to it (see Hebra). At least one eponym bears the name of a town (see Greeley). With usage come changes in the spelling of several names. Thus Saker became Suker, Türk became Tuerk, and Gradenigo has been misspelled Gradenigro. Thus, one may also find inherent errors

in this book. Though an attempt has been made to seek the most authoritative sources, there is some variation in the literature, and so the author accepts the responsibility for these inherent limitations, as well as for unintentional errors in compilation.

I would like to express my thanks to Dr. Alston Callahan for his encouragement in the preparation of this work and particularly for his suggestions for the section on eponyms in ophthalmic surgery. Also, I would like to thank my secretary, Mrs. Judi Stutts, for her invaluable assistance in the preparation of the manuscript.

It is the author's hope that this will prove a useful and reliable reference guide and a stimulus to further study.

SPENCER P. THORNTON, M.D.

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OPHTHALMIC EPONYMS

***An Encyclopedia of Named Signs,
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SECTION ONE
**SIGNS, SYNDROMES, AND DISEASES IN MEDICAL,
PEDIATRIC, AND NEURO-OPHTHALMOLOGY**

A

ACOSTA'S SYNDROME

Synonym: "*Mountain Climber's Syndrome.*"

Ocular Findings: Light adaptation defects.

Visual scintillation.

Defective color vision.

Associated Clinical Findings: Breathing coordination defects.

Muscle tonus defects.

Dysacucia.

Pathologic Etiology: Cerebral hypoxia and hyperventilation.

Bibliography

von Grósz, S.: Eponyme—Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

ADIE'S SYNDROME

Synonyms and Similar Conditions: *Holmes-Adie Syndrome, Markus Syndrome, Saenger Syndrome, Pseudo Argyll Robertson Pupil.*

Ocular Findings: Tonic pupil (pupillotonia), usually dilated, usually unilateral.

Pupil dilates and constricts SLOWLY to light and accommodation.

Pupil reacts readily to miotics and mydriatics and the dilated pupil is constricted with 2.5% mecholyl which has no visible effect on the normal pupil.

Associated Clinical Findings: Most frequent in young women.

Absent tendon reflexes (knee and ankle jerk).

Etiology: Unknown. Probable female sex linkage.

Bibliography

Adie, W.: Complete and incomplete forms of benign disorder characterized by tonic pupils and absent tendon reflexes. Brit. J. Ophthal., 16:449-461, 1932,

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Laties, A. M., and Scheie, H. G.: Adie's syndrome: Duration of methacholine sensitivity. *Arch. Ophthalm.*, 74:458-459, 1965.

ADDISON-BIERMER SYNDROME

Synonym: *Pernicious Anemia Syndrome.*

Ocular Findings: Retinal hemorrhages.
Optic atrophy.

Associated Clinical Findings: Glossitis.

Histamine refractive achylia.

May be seen in association with spinal cord tumors (Dana's syndrome).

Pernicious anemia.

Pathology: Megalocytic hyperchromic anemia.

Etiology: Vitamin B 12 or "intrinsic factor" deficiency.

Bibliography

von Grósz, S.: Eponyme—Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

ADLER'S SIGN

Synonym: *Adler's Anomaly.*

Findings: Constitutional and hereditary anomaly of the leukocytes in patients with Fuch's Gyrate Atrophy of the choroid and retina.

Pathology: Numerous azurophilic granulations (Adler's Anomaly) are seen in the polymorphonuclear leukocytes as well as in the basophiles and eosinophiles.

The presence of these azurophilic granulations is a characteristic finding in the genetic carriers of gyrate atrophy.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

ALBERS-SCHÖNBERG DISEASE

Synonyms: *Osteopetrosis, Marble Bones Disease.*

Ocular Findings: Narrowing of the optic foramina.
Optic atrophy.
Occasional progressive proptosis.

Associated Clinical Findings: Marked bony fragility and frequent fractures.
Hepatosplenomegaly.

Pathology: Increased density and thickness of cortical and spongy bone.
Blood changes resembling hyperchromic anemia or leukemia.

Etiology: Familial, may be dominant or recessive.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

ALBRIGHT'S SYNDROME

Synonyms: *Fuller-Albright Syndrome, McCune-Albright Syndrome.*

Ocular Findings: Unilateral proptosis.
Papilledema.
Optic atrophy.
Occasional visual field defects.

Associated Clinical Findings: Unilateral osteitis fibrosa cystica.
Spotty melanosis (brown skin patches).
Precocious puberty.

Etiology: Unknown. Female sex linkage reported.

Bibliography

- Albright, F.; Butler, A. M.; Hampton, A. O.; and Smith, P.: Syndrome characterized by osteitis fibrosa disseminata, areas of pigmentation and endocrine dysfunction,

with precocious puberty in females; report of 5 cases. *New Eng. J. Med.*, 216:727-746, 1937.

Hibbs, R. and Rush, H. P.: Albright's syndrome. *Ann. Intern. Med.*, 37:587-593, 1952.

ALPORT'S SYNDROME

Ocular Findings: Anterior lenticonus with abnormal thinning of the capsule.

Punctate subcapsular (sometimes described as stellate) cataract at the anterior or posterior lens pole.

Occasional microspherophakia.

Associated Clinical Findings: Familial hemorrhagic nephritis.

Deafness.

Males mostly affected.

Patients usually die before reaching the age of thirty.

Etiology: Familial. Male sex linked. Dominant autosomal.

Bibliography

Brownell, R. D. and Wolter, J. R.: Anterior lenticonus in familial hemorrhagic nephritis. Demonstration of lens pathology. *Arch. Ophthalm.*, 71:481-483, 1964.

Perrin, D.: Alport's syndrome. (Hereditary nephropathy accompanied by deafness and ocular changes.) *Ann. Oculist.*, 197:329-346, 1964.

ALSTRÖM-HALLGREN SYNDROME

Similar Condition: *Laurence-Moon-Bardet-Biedl Syndrome.*

Ocular Findings: Retinitis pigmentosa.

Defective vision.

Associated Clinical Findings: Obesity.

Diabetes mellitus.

Deafness.

Psychosis is frequent.

Etiology: Familial.

Bibliography

von Grósz, S.: Eponyme—Syndrome. An Autorennamen gebundene augenärztliche

Symptomenkomplexe und Krankheiten. Klin. Monatsbl., Augenheilk., 148:1-45, Feb., 1966.

ALZHEIMER'S DISEASE

Synonym: *Presenile Dementia*.

Ocular Findings: Agnosia.

Alexia.

Ocular fundi are not abnormal. The ocular symptoms are due to involvement of higher visual pathways.

Associated Clinical Findings: Memory loss.

Epileptiform seizures.

Spasticity.

Usually becomes manifest in middle life.

Pathology: Presenile cerebral degeneration. Characteristic findings are gliosis with glial plaques in the temporal and parietal lobes with atrophy of the ganglion cells.

Etiology: Unknown.

Bibliography

Pearlman, M. D.: *Eye Signs in Systemic Disorders*. Charles C Thomas, Springfield, Ill., 1965.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams & Wilkins, Baltimore, Md., 1957.

AMALRIC'S SYNDROME

Ocular Findings: Macular dystrophy.

Central visual defects.

Associated Clinical Findings: Deafness.

Etiology: Familial.

Bibliography

von Grósz, S.: Eponyme—Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl., Augenheilk., 148:1-45, Feb., 1966.

ANDOFSKI'S SYNDROME

Synonym: *Atopic Cataract Syndrome.*

Ocular Findings: Atopic cataract.

Associated Clinical Findings: Atopic dermatitis, a chronic superficial inflammation of the skin characterized by thickening, excoriation, and lichenification, associated with other allergies.

Pruritis which is worse at night.

Usually seen in childhood.

Etiology: Familial. Usually there is a strong family history of allergic diseases.

ANGELUCCI'S SYNDROME

Ocular Findings: Vernal conjunctivitis.

Itching, tearing, photophobia, conjunctival injection and a tenacious mucoid discharge are characteristic.

Symptoms usually recur in the spring and last through the summer.

Associated Clinical Findings: Vasomotor lability.

Emotional instability.

A history of numerous allergies is common.

Pathology: Smears of the conjunctival discharge contain numerous eosinophiles.

Etiology: Felt to be an allergic syndrome. Probably familial.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

LaGrange, H.: Pathogenic problem of so-called critical allergic conjunctivitis; specific sensitization—non-specific sensitization—instability of organic colloids. *Brit. J. Ophthal.*, 19:241-253, 1935.

ANTON-REDLICH PHENOMENON

Synonym: *Anosognosia.*

Ocular Findings: Condition in which a patient who is blind or has a homonymous hemianopia is not aware of the defect and acts as

if it did not exist. He may even describe objects that he actually does not see.

Etiology: Some feel this may be a mental disturbance.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

ANTON'S SYNDROME

Synonyms and Similar Conditions: *Denial-visual Hallucination Syndrome, see Schilder's Disease.*

Ocular Findings: Blindness.

Patient seems unaware of and undisturbed by his blindness and attempts to walk and grasp things though he is able to see nothing.

Normal pupillary reaction to light and accommodation.

Normal motility.

Occasional bilateral hemianopia.

Associated Clinical Findings: Patient gives statements of visual experiences though blind.

Intellectual deterioration, especially memory.

Tendency to confabulate.

Amnesic aphasia.

Pathology: Cortical visual loss in the sensory visual area with interference with the efferent reflexes passing to the thalamus and the ocular muscle nuclei.

Bibliography

Raney, A. A. and Nielsen, J. M.: Denial of blindness (Anton's symptom); two clinical cases. *Bull. Los Angeles Neurol. Soc.*, 7:150-151, 1942.

APERT'S SYNDROME

Synonym: *Apert-Hallerman-Streiff Syndrome.*

Ocular Findings: Exophthalmos due to shallow orbits.

Optic atrophy and consequent visual loss.

Medulated nerve fibers in the posterior pole.

Ptosis.

Strabismus.

Nystagmus.

Exposure keratitis when exophthalmos is marked.

Occasional cataracts.

Hypertelorism.

Associated Clinical Findings: Oxycephaly (tower skull).

Syndactyly.

Low mentality.

May have loss of smell.

May have loss of hearing.

Headaches in older patients.

Convulsions in severe cases.

Etiology: Congenital, familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Mann, I.: Theory of embryology of oxycephaly. *Trans. Ophthal. Soc. UK*, 55:270-300, 1935.

ARGYLL ROBERTSON PUPIL

Synonyms: *Spinal Miosis, Argyll Robertson Syndrome.*

Ocular Findings: The pupil fails to react directly or consensually to light but contracts to convergence and accommodation.

The condition is usually bilateral.

The pupil is usually miotic and irregular.

Pupil reacts poorly to cycloplegics and mydriatics.

Etiology: Uncertain but involves sympathetic interruption in or near the midbrain, especially in the region of the tectum.

Has been frequently associated with lues, spinal tumors, multiple sclerosis, encephalitis, etc.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Lowenstein, O.: Argyll Robertson pupillary syndrome; mechanism and localization. *Amer. J. Ophthal.*, 42:105-121, 1956.

ARLT'S LINES

Ocular Findings: Longitudinal fine bluish scars or striae appearing especially on the upper tarsus in the cicatricial stage of trachoma. These lines appear in contrast to the healing conjunctiva which appears smooth and grayish white. They represent subconjunctival scar tissue, and may be a precursor to entropion.

ARLT'S TRIANGLE

Similar Conditions: *See Ehrlich's Line, Krukenberg Spindle.*

Ocular Findings: A triangle of fine pigmented KPs on the endothelium of the inferior cornea. Because of the convection currents of the anterior chamber, the base of the triangle is down.

There is usually a brief clear area between the base of the triangle and the limbus.

Etiology: This pattern of KPs, like most other endothelial deposits of this type, is a sign of low grade uveitis.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

ARNOLD-CHIARI SYNDROME

Synonyms: *Arnold-Chiari Deformity, Platybasia.*

Ocular Findings: Oculomotor palsies.

Nystagmus, especially vertical.

Eyegrounds usually normal but may show papilledema.

Occasional diplopia.

Associated Clinical Findings: Hydrocephalus.

Cerebellar ataxia.

Bilateral pyramidal tract involvement.

Pathology: Herniation of cerebellar tissue into the foramen magnum with consequent brain-stem compression.

X-ray examination shows platybasia and narrowing of the foramen magnum.

Bibliography

Cogan, D. G. and Barrows, L. J.: Platybasia and Arnold-Chiari malformation. *Arch. Ophthalm.*, 52:13-29, 1954.

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

ARNOLD-PICK SYNDROME

Synonyms: *Aphasia-agnosia-apraxia Syndrome, Pick's Disease.*

Ocular Findings: Apperceptive blindness.

Normal eyegrounds.

Inability to fix reflexly on objects within area of gaze.

Associated Clinical Findings: Progressive dementia.

Aphasia of motor type.

Apraxia.

Agnosia.

Apathy.

Occurs more in females aged 40 to 70.

Pathology: Blindness is caused by a widespread lesion involving the occipital lobe.

Electroencephalography demonstrates cerebral atrophy.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

Hassin, G. B. and Levitin, D.: Pick's disease; clinicopathologic study and report of case. *Arch. Neurol. Psychiat.*, 45:814-833, 1941.

ARRUGA'S SIGN

Ocular Findings: Bleeding on corneo-limbal incision. Leads to early operative hyphema.

Pathology: This has been described as a sign of the presence of heterochromic cyclitis or uveitis or other sympathetic diseases with ocular involvement.

ASCHER'S SYNDROME

Similar Condition: *Laffer-Ascher Syndrome.*

Ocular Findings: Swollen lids (blepharochalasis).
Spastic entropion.

Upper lids may be flaccid with redundant skin folds due to elastorhexis.

Associated Clinical Findings: Cleft upper lip.
Non-toxic goiter.

Pathology: Atrophy of the dermis and secondary relaxing of the subcutaneous tissues.

Etiology: Familial. Dominant autosomal.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

AUBINEAU-LENOBLE SYNDROME

Synonyms: *Lenoble-Aubineau Syndrome, Nystagmus-myoclonia Syndrome.*

Ocular Findings: Congenital nystagmus.

Associated Clinical Findings: Fasciculations of the muscles elicited by cold or mechanical stimulation.

Tremors of the head and limbs.

Vasomotor disturbances.

Hyperreflexia.

Pathology: Diffuse non-specific meningovascular and glial changes characteristic of chronic toxemia.

Etiology: Hereditary. Recessive familial.

Bibliography

Lenoble, R.: *Rev. de Med., Paris*, 26:471, 1906.

AVELLIS SYNDROME

Ocular Findings: Horner's Syndrome (miosis, ptosis and relative enophthalmos).

Associated Clinical Findings: Paralysis of the soft palate.

Paralysis of the vocal cords.

Occasional loss of pain and temperature sense.

Anhidrosis on the side of Horner's.

Pathology: Lesion implicating the nucleus ambiguus and involving the descending sympathetics.

Occurs most frequently as the result of vascular or inflammatory lesions but may be caused by neoplasms or trauma.

Bibliography

Fox, S. L. and West, G. B., Jr.: Syndrome of Avellis; review of literature and report of one case. *Arch. Otolaryng.*, 46:773-778, 1947.

AXENFELD-SCHÜREMBERG SYNDROME

Ocular Findings: Cyclic oculomotor paralysis. Paralysis of the extraocular muscles alternating with spasm. Condition is congenital.

Etiology: Familial.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

Latorra Morasso, L. S. and Aguilar, J.: Axenfeld-Schüremberg disease. *Amer. J. Ophthalm.*, 27:1172, 1944. (Abs.)

AXENFELD'S SYNDROME

Synonyms: *Posterior Embryontoxon, Irido-corneal Dysgenesis.*

Ocular Findings: Circumcorneal gray opacity deep in the stroma extending several millimeters out from the limbus. Appears like embryontoxon.

Associated with anomalies of the iris and anterior chamber angle.

May have polycoria, corectopia, dyscoria, ectropion uveae.

May have congenital corneal opacities, iris processes adherent to Schwalbe's Ring.

May have ectopia lentis.

Usually is present in early childhood or adolescence.

Results in a type of juvenile glaucoma.

Pathology: Irido-corneal dysgenesis with a prominent anterior ring of Schwalbe.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Sugar, H. S.: Juvenile glaucoma with Axenfeld's syndrome. A histologic report. *Amer. J. Ophthal.*, 59:1012-1019, 1965.

B

BABINSKI-NAGEOTTE SYNDROME

Ocular Findings: Horner's Syndrome (ptosis, miosis and enophthalmos) on the side of the lesion.

Nystagmus.

Associated Clinical Findings: Cerebellar hemiataxia on the side of the lesion.

Hemiparesis on the contralateral side.

Disturbance of sensibilities on the contralateral side.

Pathology: Lesion (usually vascular or neoplastic) in the ponto-bulbar or medullopontine transitional region.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

BAER'S NYSTAGMUS

Synonym: *Reflex Nystagmus*.

Ocular Findings: A fine nystagmus of very high frequency which may be seen in erosions or other superficial lesions of the cornea. It

has not been adequately demonstrated whether or not it consists of regular or irregular eye movements.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examinations*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

BALINT'S SYNDROME

Ocular Findings: Psychic paralysis of fixation.

Fixation is in abeyance, but the eyes can be moved freely in any direction.

Optic ataxia.

Disturbances of visual attention.

Pathology: Post oculogyric apparatus lesions. Due to bilateral lesions involving the occipito-parietal cortex.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Hecaen, H. and de Ajuriaguerra, J.: Balint's syndrome (psychic paralysis of visual fixations) and its minor forms. *Brain*, 77:373-400, 1954.

BALLET'S DISEASE

Synonym: *Ballet's Sign (in hyperthyroidism).*

Ocular Findings: Ophthalmoplegia externa. Specifically, impairment of ocular movements without internal ophthalmoplegia.

Associated Clinical Findings: Hyperthyroidism or Graves' Disease.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examinations*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

BANCROFT'S FILARIASIS

Ocular Findings: Elephantiasis of the eyelids.

Iritis.

Retinal hemorrhages.

Associated Clinical Findings: Bancroft's filariasis is seen principally in Australia. It is caused by a nematode parasite transmitted by insect vector.

Pathology: The ocular manifestations are caused by the actual presence of the worm in the tissues. In the case of iritis, the adult worm has been detected in the anterior chamber. Occasionally, the microfilariae may be present in the secretions of the lacrimal glands.

Bibliography

Toussaint, D. and Danis, P.: Retinopathy in generalized loa-loa filariasis. A clinico-pathological study. Arch. Ophthal., 74:470-476, 1965.

BARDET-BIEDL SYNDROME

Synonym or Similar Condition: *Laurence-Moon-Biedl Syndrome.*

Ocular Findings: Diminished vision.

Tapeto-retinal degeneration; may appear as classic pigmentary retinopathy or occasionally as atypical pigmentary retinopathy.

May have macular degeneration.

May have hemeralopia.

May have nystagmus.

Associated Clinical Findings: Obesity of Frolich's type.

Genital dystrophy or hypoplasia.

Polydactyly.

Mental deficiency.

Etiology: Hereditary.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BARRÉ-LIEOU SYNDROME

Synonym: *Posterior Cervical Sympathetic Syndrome.*

Ocular Findings: Pain in the eyes.

Transitory amaurosis.

Hypesthesia of the cornea.

Recurrent trophic ulcers of the cornea confined to the palpebral fissure area.

Associated Clinical Findings: Headache.

Vertigo.

Tinnitus.

Vasomotor disturbances involving the face.

Arthritis is frequently associated with this syndrome.

Pathologic Etiology: Usually caused by irritation or trauma to the vertebral nerve with resultant disturbance of circulation to the cranial nuclei, especially the 5th and 8th cranial nerves.

Bibliography

Durham, R.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Pearlman, M. D.: *Eye Signs in Systemic Disorders*. Charles C Thomas, Springfield, Ill., 1965.

BARTENWERFER'S SYNDROME

Ocular Findings: Epicanthus.

Mongoloid eyelid fissures.

Blepharophimosis.

Hypertelorism.

Associated Clinical Findings: Short stature (retarded growth).

Congenital dislocation of the hip is frequently associated.

Spinal Curvature.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

BASEDOW'S DISEASE

Synonyms: *Grave's Disease, Exophthalmic Goiter, Parry's Disease, Thyrotoxicosis.*

Ocular Findings: Proptosis.

Asymmetry of the eyes.

Thickening of the upper lids.
On upward gaze the lids may move faster than the eyes.
Jerky lateral gaze movements.
Absent forehead wrinkles on upward gaze.
See Grave's Disease for more complete listing of ocular signs.

Associated Clinical Findings: Systemic signs of hyperthyroidism.

Tachycardia.
Trembling.
Emaciation in severe cases.
Increased basal metabolism.
Women are usually affected more than men.

Etiology: Exact etiology unknown, but a strong familial tendency to the disease has been observed.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill. 1956.

BASEDOW'S SIGN

Ocular Findings: Reduced orbital resiliency in congestive dysthyroid exophthalmos.

Orbital resiliency is normal in non-congestive exophthalmos.
It may be determined by applying pressure against the upper and lower lids with the eye open so that movements of the globe can be observed grossly.

In non-congestive exophthalmos, the eye may be pushed back into the orbit several millimeters. In congestive exophthalmos, resistance is increased.

This sign precedes all other signs of congestion.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

BASSEN-KORNZWEIG SYNDROME

Ocular Findings: Retinitis Pigmentosa.

Associated Clinical Findings: Signs of spino-cerebellar degeneration.

Celiac Disease.
Signs overlap those of Refsum's Syndrome.

Pathology: A "spiny" appearing erythrocyte (acanthocyte) is seen on blood smear. Serum beta-lipoprotein is absent.

Etiology: In Bassen-Kornzweig Syndrome, the deficiency of beta-lipoprotein lowers all serum fats, including the fat soluble vitamin A, leading to a degeneration of the visual cells.

Bibliography

- Bassen, F. A. and Kornzweig, A. L.: Malformation of the erythrocytes in cases of atypical retinitis pigmentosa. *Blood*, 5:381-387, 1950.
 Baum, J. L.; Tannenbaum, M.; and Kolodny, E. H.: Refsum's syndrome with corneal involvement. *Amer. J. Ophthalm.*, 60:699-708, 1965.
 Gills, J. P.; Hobson, R.; Hanley, W. B.; and McKusick, V. A.: Electro-retinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. *Arch. Ophthalm.*, 74:596-603, 1965.

BATTEN-MAYOU SYNDROME

Synonyms: *Batten's Disease, Vogt-Spielmeyer Syndrome, Juvenile Amaurotic Family Idiocy.*

Ocular Findings: Macular degeneration.

Loss of the foveal reflex.

Diffuse pigmentary disturbance of the retina.

The optic nerve eventually becomes atrophic.

Retinal vessels are reduced in caliber.

May look like retinitis pigmentosa.

Occasionally may see a cherry red spot in the macula.

Pathology and Clinical Findings: Juvenile amaurotic family idiocy usually becomes clinically manifest at puberty or in the late teens. It differs from the infantile form in that the rapidly progressing degeneration of ganglion cells does not occur; therefore the cherry red spot in the macula is usually not seen.

Etiology: Familial. Simple recessive inheritance.

Bibliography

- Hoffman, J.: Pigmentary retinal lipid neuronal heredodegeneration (Spielmeyer-Vogt disease): neuro-ophthalmologic considerations. *Amer. J. Ophthalm.*, 42:15-21, 1956.
 Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

BATTEN'S MYOTONIC DYSTROPHY

Synonyms: *Steinert's Disease, Batten-Steinert Syndrome.*

Ocular Findings: Ptosis.

Cataracts, usually posterior subcapsular, beginning in the second decade of life.

Trophic keratitis.

Conjunctivitis.

Associated Clinical Findings: Myopathies of the face, especially of the chewing muscles.

Weakness of the muscles of the arms and thighs.

Precocious baldness.

Testicular atrophy.

Emotional lability and emotional disturbances.

Etiology: Familial. Dominant inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BEAL'S CONJUNCTIVITIS

Synonym: *Pharyngoconjunctival Fever.*

Ocular Findings: Acute follicular conjunctivitis.

Transient keratitis.

Lacrimation.

Clinically identical with inclusion conjunctivitis but without inclusions.

Heals spontaneously without corneal damage.

Associated Clinical Findings: Fever in about 90% of cases.

Preauricular adenopathy.

Pharyngitis in most cases.

Most common in children and young adults.

Incubation period from five to seven days.

Subsides in two to three weeks.

Etiology: Generally thought to be due to adenovirus type three.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
- Theodore, F. H.: Pharyngoconjunctival fever and epidemic keratoconjunctivitis. *Eye Ear Nose Throat Monthly*, 45:94-95, 1966.

BEHÇET'S SYNDROME

Ocular Findings: Recurrent uveitis, or iridocyclitis.

Hypopyon during acute stages.

May have visual loss.

May have retinal hemorrhages.

Acute stages may be accompanied by nystagmus.

Associated Clinical Findings: Oral aphthous ulcers.

Genital aphthous ulcers.

Condition is chronic with remissions and exacerbations.

Acute episodes regress spontaneously, but complete remission is rare.

Occurs mainly in adult males.

Found primarily in the Middle East and in the Mediterranean area.

Etiology: Uncertain but thought to be viral.

Bibliography

- Fenton, R. H. and Eason, H. A.: Behcet's syndrome. *Arch. Ophthalmol.*, 72:71-80, 1964.
- Gray, G. S.: Triple complex syndrome of Behcet. *Canad. M. A. J.*, 62:597-599, 1950.
- Mamo, J. G. and Bogldassarian, A.: Behcet's disease. *Arch. Ophthalmol.*, 71:4-13, 1964.

BEHR'S ABDUCTION PHENOMENON

Ocular Findings: The abducting eye shows *miosis* on conjugate lateral gaze when looking toward the side of a sixth nerve palsy. The eye may otherwise show general rigidity or light rigidity of the pupil.

Behr's abduction phenomenon is thus opposite to Tournay's.

Etiology: Usually luetic.

Bibliography

- Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

BEHR'S DISEASE

Synonym: *Central Tapeto-retinal Degeneration.*

Ocular Findings: Appearance of a central atrophic chorioretinitis, frequently accompanied by peripheral retinal lesions of the pigmentary degenerative type.

Clinical Pathology: Felt to be an adult or pre-senile form of cystic macular degeneration. Usually becomes clinically apparent in the third, fourth or fifth decades.

Etiology: Familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BEHR'S SIGN

Synonym: *Hemianopic Tract Sign.*

Ocular Findings: Anisocoria associated with a hemianopia with the pupil dilated but normally reactive on the side contralateral to the involved optic tract.

In a lesion of the right optic tract, causing left sided hemianopia, the stimulus that reaches the nucleus of the left pupillary sphincter would be diminished, thus resulting in a wider left pupil.

Etiology: Behr suggested that each optic tract is in more intimate relation with the contralateral sphincter nucleus than with the homolateral one.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Cogan, D. C.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

BEHR'S SYNDROME

Synonym: *Optic Atrophy-ataxia Syndrome.*

Ocular Findings: Bilateral atrophy of the optic nerve, more severe temporally.

Infantile amaurosis.

Nystagmus.

Associated Clinical Findings: Pyramidal tract signs characterized by increased tendon reflexes and positive Babinski.

Disturbance of coordination with ataxia.

Mental retardation of varying degrees.

Onset in infancy.

Etiology: Thought to be an infantile form of heredofamilial optic atrophy and hereditary ataxia.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Franceschetti, A.: Le syndrome de Behr, ses rapports avec la maladie de Leber et les hérédoataxies. *Ophthalmologica*, 107:17-25, 1944.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BELL'S PALSY

Synonym: *Bell's Paralysis.*

Ocular Findings: Palpebral fissure is wider on the affected side.

Eyelids cannot be closed on the affected side.

Epiphora may be seen because of eversion of the lower lid.

Winking is impaired on the involved side.

Associated Clinical Findings: Paralysis of the facial nerve following inflammation in the stylomastoid foramen.

Occasional pain in the side of the face.

Forehead is smooth and the eyebrow is somewhat lower than on the sound side.

The patient cannot frown.

Occur more in young adult males, but can affect anyone at any age.

Most recover gradually in a few months.

Pathology: Degeneration of the medullary sheath and axis cylinder of the involved nerve has been reported.

Etiology: No definite etiologic agent has been confirmed.

Bibliography

Reddy, J. B. and Balshi, S.: Histopathology of Bell's palsy. *Eye Ear and Nose Throat Monthly*, 45:62-65, 1966.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

BELL'S PHENOMENON

Ocular Findings: Commonly understood as the normally found upward and outward turning of the eyes on firm closure of the lids, produced by the reflex association of the superior rectus with the orbicularis muscle.

But also described as an elevation of the lids not only with the normal voluntary upward movements of the eyes but also with involuntary upward movements on attempted closure of the lids. In other words, the lid opens widely when an attempt is made to close it (Cogan, page 73).

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

BENEDIKT'S SYNDROME

Synonym: *Tegmental Syndrome*.

Ocular Findings: Homolateral partial third nerve paralysis.
External ophthalmoplegia, homolateral.

Associated Clinical Findings: Intention tremor of the contralateral arm and leg.

Paresis of the contralateral extremities if the pyramidal tract is involved.

Pathology: Seen in cases with lesion involving the third nerve nucleus and affecting the red tegmental nucleus.

Bibliography

- Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.
 Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

BENJAMIN'S SYNDROME

Ocular Findings: Epicanthus.

Associated Clinical Findings: Enlarged skull.

Oligophrenia.

Constitutional anemia (hypochromic with normal WBC).

Premature birth is common.

Etiology: Congenital.

Bibliography

- von Grósz, S.: Eponyme—Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

BENSON'S DISEASE

Synonym: *Asteroid Hyalitis*.

Ocular Findings: Tiny particles of calcium soap seen suspended in the vitreous.

Usually found more frequently in older adults in the 50 plus age group.

Vision is not usually affected.

Associated Clinical Findings: More common in males; three to one over females.

Relatively high percentage of cases have been reported in association with diabetes (60% to 70%).

Most cases are felt to be not clinically significant.

Bibliography

- Rutherford, C. W.: Asteroid bodies in vitreous. *A.M.A. Arch. Ophthal.*, 9:106-117, 1933.

BERGMEISTER'S PAPILLA

Ocular Findings: A conical shaped mass of glial elements trapped in the center of the optic cup by growing nerve fibers. It appears white to gray and forms a pre-retinal or papillary veil.
May project several diopters in front of the optic disc.
Does not affect vision.

Associated Clinical Findings: None. Of no clinical significance.

Etiology: A congenital anomaly.

Bibliography

Hogan, M. J., and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

BERLIN'S DISEASE

Synonyms: *Berlin's Macular Edema, Commotio Retinae.*

Ocular Findings: Traumatic edema of the retina, especially the macula. Usually seen following blunt contusion of the eye.
Not infrequently followed by subsequent retinal detachment or macular hole.

Pathology: The percipient elements of the fovea are destroyed resulting frequently in a permanent central macular atrophy and consequent scotoma.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology, 6th ed.*, W. B. Saunders, Philadelphia, Pa., 1959.

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

BERNARD-HORNER SYNDROME

Synonym: *Horner's Syndrome.*

Ocular Findings: Miosis.
Ptosis or narrow palpebral fissure.

Relative enophthalmos.

Pupil is usually constricted but reacts to light and accommodation and there is retention of the lid reflex. The pupil fails to dilate with cocaine, but will dilate with adrenalin, which differentiates it from a spastic miosis.

Associated Clinical Findings: Ipsilateral anhidrosis of the face and neck.
Occasional facial hemiatrophy.

Pathology: Interruption of the sympathetic chain in the neck.

Bibliography

Horner, F.: Ueber eine form von ptosis. *Klin. Mbl. Augenheilk.*, 7:193-198, 1869.

BESNIER-BOECK-SCHAUMANN SYNDROME

Synonyms and Similar Conditions: *Schaumann's Disease, Boeck's Sarcoidosis.*

Ocular Findings: Granulomatous uveitis, with large mutton-fat KPs, iris nodules, and flare in the anterior chamber.

May have mild to severe posterior uveitis.

May have vitreous floaters.

May be complicated by exudative retinitis.

May have band keratopathy.

Associated Clinical Findings: Lymphadenopathy.

Skin rash of upper parts of body.

Enlarged Hilar nodes may be seen on x-ray.

Lethargy and easy fatiguability.

Onset usually in the third and fourth decades.

Pathology: Granulomatous lesions found in the involved tissues.

Etiology: Unknown, though a racial predilection for the Negro is known.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

BEST'S MACULAR DEGENERATION

Ocular Findings: Congenital macular degeneration.

Appears in early childhood.

Decreased central acuity.

Pathologic Etiology: Heredo-familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BIBER-HAAB-DIMMER CORNEAL DYSTROPHY

Synonym: *Lattice Dystrophy*.

Ocular Findings: Lattice like dystrophy of the cornea.

Diffuse involvement of the stroma, unilateral or bilateral.

Stroma shows hyaline degeneration and absence of acid mucopolysaccharides (as in granular dystrophy, but contrasted with macular dystrophy).

Etiology: Familial, recessive.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. *Arch. Ophthalmol.*, 74:517-520, 1965.

BICKEL-BING-HARBOE SYNDROME

Ocular Findings: Neuritis of the optic nerve.

Central visual defects.

Associated Clinical Findings: Neuroses and psychic changes.

Hyperglobulinemia.

Etiology: Unknown.

Bibliography

von Grósz, C.: Eponyme-Syndrome. An Autore namen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

BIDWELL'S GHOST

Synonym: *Purkinje After-image.*

Ocular Findings: Negative after images with qualities opposite those of the original stimulus.

They are produced by a modified second neural stimulus and are called "induced" or "second" after-images. For instance, if one looks at a white light, the negative after image, or Bidwell's ghost, will appear dark.

See Hering's After-image.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1956.

BIELSCHOWSKY-JANSKY SYNDROME

Synonyms and Similar Conditions: *Batten-Mayou Syndrome, Vogt-Spielmeyer Syndrome, Juvenile Amaurotic Idiocy, Stock-Spielmeyer Syndrome.*

Ocular Findings: Macular degeneration and loss of foveal reflex.

Diffuse pigmentary disturbance of the retina.

May look like retinitis pigmentosa.

Marked visual loss, progressing to amaurosis.

Usually occurs at puberty or late teens.

It differs from the infantile form in that the cherry-red macula is not usually seen.

Associated Clinical Findings: Marked mental retardation.

Epileptiform seizures may be seen.

Ataxias may be seen.

Etiology: Familial. Autosomal recessive.

Bibliography

Hoffman, J.: Pigmentary retinal lipid neuronal heredodegeneration (Spielmeyer-Vogt disease); neuro-ophthalmologic considerations. Amer. J. Ophthal., 42:15-21, 1956.

BIELSCHOWSKY-LUTZ-COGAN SYNDROME

Synonym: *Internuclear Ophthalmoplegia.*

Ocular Findings: Unilateral or bilateral palsy of the internal rectus muscle during conjugate lateral movements, with normal function of this muscle in accommodation and convergence.

Nystagmus which appears in the maximally abducted contralateral eye.

Pathologic Etiology: A supranuclear palsy. Anatomically it is caused by an interruption of the medial longitudinal fasciculus connecting the nuclei of cranial nerves III, IV, and VI, especially those fibers coordinating conjugate lateral movement.

Bibliography

Orlowski, W. J.; Slomski, P.; and Wojtowicz, S.: Bielschowsky-Lutz-Cogan syndrome. *Amer. J. Ophthalm.*, 59:416-428, 1965.

BIELSCHOWSKY'S PHENOMENON

Synonym: *Doll-head Phenomenon.*

Ocular Findings: In gaze palsy, the patient cannot perform *volitional* movement to one side, but on turning the head away from that side, the eyes will make a compensatory fixating movement toward the paretic side.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md. 1957.

BIELSCHOWSKY'S PHENOMENON

Synonym: *Occlusion Hypertropia.*

Ocular Findings: In persons with so called alternating hypertropia, darkening or occlusion of the field of the fixating eye results in a downward movement of the non-fixating eye.

The patient's eyes are dissociated by holding a hand in front of one

eye. A graded occluder is placed in front of the fixating eye and the non-fixating eye is observed behind the hand of the examiner. As the field of the fixating eye is progressively darkened, the hypertropic, non-fixating eye moves down toward or even below the level of the fixating eye.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

BIELSCHOWSKY'S SIGN

Ocular Findings: In paresis of the superior oblique muscle, on having the head tilted to the involved side, the patient gets hypertropia and diplopia.

Bibliography

Berke, R. N.: *Strabismus*. (chap.) C. V. Mosby, St. Louis, Mo., 1962.

BIELSCHOWSKY'S SYNDROME

Ocular Findings: Dissociated vertical divergence.

Loss of vertical comitancy due to suppression of fusion.

May present as a phoria or tropia.

When the elevating center of the eyes are unable to work harmoniously, a temporary loss of vertical concomitancy of the eyes occurs.

Pathologic Etiology: This condition may be produced by any means which causes a suppression of fusion, such as suppression amblyopia, accommodative disturbances, etc.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

BIEMOND SYNDROME

Similar Conditions: *Bardet-Biedl*, see also *Laurence-Moon-Biedl*.

Ocular Findings: Colobomas of the iris.

May have atypical pigmentary retinopathy of the retina.

Associated Clinical Findings: Hypophyseal infantilism.

Genital hypoplasia.

Mental retardation.

Polydactyly.

Etiology: Familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BIETTI'S CORNEAL DYSTROPHY

Synonym: *Marginal Crystalline Dystrophy*.

Ocular Findings: Scintillating punctate opacities in the superficial layers of the corneal stroma in the region of the limbus.

Etiology: Familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

BITOT'S SPOT

Ocular Findings: Appears as a foamy, dry, wedge-shaped area on the bulbar conjunctiva with the base at the limbus and the apex extending toward the lateral canthus.

Other signs of xerosis may be present.

Nyctalopia may be associated.

May be complicated by keratomalacia, if not treated.

Pathologic Etiology: Keratinization of the conjunctiva due to Vitamin A deficiency.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

BJERRUM SCOTOMA

Synonym: *Arcuate Scotoma.*

Ocular Findings: The Bjerrum scotoma is a visual field defect characteristic of glaucoma. It is a nerve fiber bundle defect extending from the blind spot, sweeping around the macular region, and ending in a straight line on the nasal side corresponding to the temporal raphe in the retina.

The Bjerrum scotoma may be above, below, or both, but is more generally found below.

When the Bjerrum scotoma occurs both above and below the point of fixation, they meet in the nasal field on the raphe. The frequent discontinuity at this junction is known as the Roenne Step.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

BLAINT'S SYNDROME

Synonym: *Ocular Motor Apraxia.*

Ocular Findings: Loss of purposive eye movements.

Preservation of random eye movements.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

BLEGVAD-HAXTHAUSEN SYNDROME

Similar Condition: *Robstein's Syndrome.*

Ocular Findings: Zonular cataracts.

Associated Clinical Findings: Osteogenesis imperfecta.

Anetoderma (relaxation of the skin).

Pathology: Elevated serum alkaline phosphatase.

Etiology: Familial.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

BLESSIG CYST

Synonym: *Iwanoff Cyst*.

Ocular Findings: Peripheral cystic degeneration of the retina at the ora serrata. Occurs mainly temporally.

May predispose to retinal detachment.

May extend six or seven millimeters back from the ora.

Occurs in the non-functional area of the retina.

Usually appears after age thirty but has been reported in children.

Pathology: These cysts arise in the external plexiform layer and extend almost to the inner and outer limiting membranes. Their walls are formed of compressed glial fibers.

When these walls give way, the retina becomes split into two layers, hence the term retinoschisis.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

BLOCH-SULZBERGER SYNDROME

Similar Condition: *Naegli's Syndrome*.

Ocular Findings: Nystagmus.

Strabismus.

Optic atrophy.

Pseudoglioma may be seen.

Cataracts may be seen.

Associated Clinical Findings: Bullous eruption at birth followed by variable cutaneous pigmentations (lines and whorls).

Spotty alopecia.
Dental anomalies.

Etiology: Hereditary, familial.

Bibliography

- Duke-Elder, Sir S.: *System in Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Haber, H.: Bloch-Sulzberger syndrome (incontinentia pigmenti). *Brit. J. Derm.*, 64:129-140, 1952.

BOECK'S SARCOID

Synonym: *Besnier-Boeck Syndrome, Schaumann's Syndrome.*

Ocular Findings: Anterior granulomatous uveitis with large mutton-fat KPs and iris nodules.

May have mild to severe posterior uveitis with or without demonstrable "candle dripping" lesions.

May be complicated by glaucoma, cataracts, secondary keratitis, and optic atrophy.

Associated Clinical Findings: Rash on face and upper body.

Parotitis.

Cervical or generalized lymphadenopathy.

Reticuloendothelial involvement.

Lung disease.

More common in negroes.

Pathology: Granulomatous lesions found in the involved tissues.

Etiology: A possible auto-immune mechanism has been suggested. Racial predelection for negroid genotype.

Bibliography

- Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

BOGORAD'S SYNDROME

Synonyms: *Crocodile Tear Syndrome, Paroxysmal Lacrimation Syndrome.*

Ocular Findings: Unilateral or bilateral lacrimation while eating or drinking.

Sudden lacrimation may occur when salivation is stimulated by certain odors or the suggestion of food.

Pathologic Etiology: Thought to be due to an inter-connection of salivary nerve fibers to the lacrimal gland.

Bibliography

Russin, L. A.: Paroxysmal lacrimation during eating as sequel of facial palsy; syndrome of crocodile tears. JAMA, 113:2310-2311, 1939.

BONNET'S SYNDROME

Synonyms: *Bonnet-de-Chaume-Blanc Syndrome, Neuroretinoangiomatosis Syndrome.*

Ocular Findings: Congenital retinal angiomatosis with tortuosity of the vessels of the retina.

Arteriovenous angiomas of the optic nerve.

Associated Clinical Findings: Arteriovenous angiomas of the thalamus and the mesencephalon.

See also Sturge-Weber syndrome and von Hippel-Lindau syndrome.

Etiology: Hereditary, familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Geeraets, W. J.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

BONNEVIE-ULLRICH SYNDROME

Ocular Findings: Congenital paralytic strabismus.

Ptosis.

Hypertelorism.

Epicanthus.

Associated Clinical Findings: Various congenital somatic deformities such as regional muscular atrophy of the thorax and arms.

Deformities of the ears may be seen.

May have aplasia of body hair.

May have infantilism.

Deformities of the extremities such as clubfeet and syndactyly are frequently seen.

Etiology: Hereditary, familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BORNA DISEASE

Synonym: *Equine Encephalitis*.

Ocular Findings: Photophobia.
Nystagmus.

Associated Clinical Findings: Encephalitis.

The Eastern type is associated with about 60% mortality.

The Western type is similar to St. Louis encephalitis and has a lower mortality.

Etiology: A virus disease. Two strains of virus have been isolated, with differing associated mortality as noted above.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

BOSTON'S SIGN

Ocular Findings: Jerky lid lag on downward gaze in cases of hyperthyroidism.

Other associated lid signs of Graves' disease.

Associated Clinical Findings: Graves' disease.

Trembling, tachycardia and increased basal metabolism.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

BOURNEVILLE'S DISEASE

Synonym: *Tuberous Sclerosis*.

Ocular Findings: Retinal glial tumors.

Retinal lesions present as a mushroom-like tumor of grayish-white color in the posterior pole.

May also have small retinal hemorrhages.

May have cystic changes in the retina.

Epiloia.

Associated Clinical Findings: Adenoma sebaceum.

Mental deficiency.

Epilepsy.

May have misshaped ears.

May also have associated congenital heart and kidney defects.

Prognosis is poor.

Usually becomes apparent in childhood.

Etiology: Heredofamilial, irregular dominant transmission. More frequent in females.

Bibliography

Datelines in Ophthalmology, Vol. 2., No. 1., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Ross, A. T. and Dickerson, W. W.: Tuberous sclerosis. *Arch. Neuro. Psychiat.*, 50:233-257, 1943.

BOWEN'S DISEASE

Synonym: *Intraepithelial Epithelioma*.

Ocular Findings: Epibulbar tumor situated near the limbus.

Appears diffuse, flat to slightly elevated, vascularized and boggy to granulomatous.

Considered to be carcinoma-in-situ.

May develop into squamous cell carcinoma.

Associated Clinical Findings: A great percentage are associated with primary cancers internally or elsewhere in the skin.

Bibliography

- Donaldson, D.: Bowen's carcinoma (intraepithelial epithelioma). Arch. Ophthal., 71:712-713, 1964.
Graham, J. H. and Helwig, E. B.: Bowen's disease and its relationship to systemic cancer. Arch. Derma. Syph., 80:133-159, 1959.
Reese, A. B.: *Tumors of the Eye*, 2nd ed. Harper and Row, New York, N.Y., 1963.

BRACHMAN-deLANGE SYNDROME

Synonym: *Typus Ametelodamensis*, see de Lange.

Ocular Findings: Bushy eyebrows that are fused in the midline.
Eyelashes are long and upturned.

Associated Clinical Findings: Nose is small with antiverted nostrils.
Distance between the nose and upper lip is wide.
Corners of the mouth are turned downward giving a crescent shaped appearance.
The skull is brachycephalic and small.
The mandible is usually small.

Etiology: Congenital.

Bibliography

- Keitel, H.: Lippincott's Med. Sci., 17:47, 1966.
Opitz, J.: *Birth Defects Reprint Series*, 22, 1965.

BRAILSFORD'S SYNDROME

Synonym: *Morquio's Disease*.

Ocular Findings: Fine diffuse haze throughout the corneal stroma. Bilateral.
Hypertelorism.
Vision is not affected.

Associated Clinical Findings: Familial chondro-osteodystrophy.
Dwarfism.

Large head.
Flexion difficulties.
Mentality normal.

Pathology: Laboratory findings of mucopolysaccharidosis.

Etiology: Familial. Autosomal recessive.

Bibliography:

Gills, J. P.; Hobson, R.; and Hanley, W. B., *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. Arch. Ophthalm., 74:596-603, 1965.

BRILL'S DISEASE

Synonym: *Typhus Fever.*

Ocular Findings: Hyperemia of the conjunctiva appearing before onset of fever.

Occasional conjunctival hemorrhages.

Photophobia.

Miosis in acute stages.

Optic neuritis (often bilateral) is frequent.

Cataract may result from intraocular inflammation.

Associated Clinical Findings: Acute onset of headache, chills and fever and vomiting.

Rash in the axillary and flank regions appearing about four days after onset; gradually becoming generalized.

The majority of cases have central nervous system involvement with delirium tremens, tremors, and hemiparesis.

Etiology: Louse and flea borne. Caused by *R. prowazeki*.
Incubation period seven to twenty-one days.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

BROOK'S DISEASE

Synonym: *Cystic Adenoid Epithelioma*.

Ocular Findings: Sebaceous adenomas of the eyelids.

It is characterized by the presence of numerous small symmetrical tumors of a rosy-yellow color, having the appearance of pearls or sebaceous adenomas.

They tend to be localized on the face more particularly around the eyelids, extending sometimes to the scalp and to the seborrheic regions of the trunk.

Pathology: Brooks' disease is a case of benign proliferation of the basal cells, often in connection with the hair follicles.

Etiology: Familial. Dominant transmission.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BROWN-SEQUARD SYNDROME

Ocular Findings: Nystagmus present in about 10% of cases, with high cervical cord lesions.

Associated Clinical Findings: Motor paralysis and loss of deep tendon and vibratory sensation on one side.

Loss of pain and temperature sense on the opposite side.

Clinically, the syndrome is rarely seen in its complete form.

The neurologic findings are variable depending on the degree of involvement of the tracts of the cord.

Pathologic Etiology: Due to spinal cord tumors, or in certain cases, trauma.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Sloan, L. H.: Clinical syndrome manifested by various types of compression of spinal cord: differential diagnosis of spinal cord tumors. *Surg. Clin. N. Amer.*, 17:559-577, 1937.

BROWN'S SHEATH SYNDROME

Synonym: *Brown's Superior Oblique Tendon Sheath Syndrome.*

Ocular Findings: Restriction of elevation in the nasal field with no overaction of the superior oblique.

May be accompanied by *underaction* of the inferior oblique with a widening of the palpebral fissure on attempted elevation.

The clinical picture gives an impression of a superior oblique palsy of one or both eyes, because the eyeballs do not move in the direction of their muscles' action.

The testing of the passive motility of the eyeballs has a decisive diagnostic importance.

The lack of passive movements of the eye in the direction of the inferior oblique action speaks for the Brown Syndrome.

Pathologic Etiology: Caused by the inhibiting action of the fibrosed and shortened sheath of the superior oblique tendon of the ipsilateral eye. Effect due to sheath attachment at the trochlea.

Accurate diagnosis can only be confirmed by forced ductions at surgery.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Krzystkova, K.: The Brown syndrome, the syndrome of the superior oblique tendon sheath. *Klin. Oczna.*, 34:169-172, 1964.

BRUNS'S SYNDROME

Similar Condition: *See Nothnagel's Syndrome.*

Ocular Findings: Partial third nerve paralysis.

Gaze pareses combined with some degree of internal or external ophthalmoplegia.

May have ptosis of the upper lids, uncommon.

Associated Clinical Findings: Trunkal ataxia.

Ataxia appears first, then followed by bilateral oculomotor involvement.

Pathologic Etiology: Caused by a lesion in the cerebellum.

Contrasted with Nothnagel's Syndrome in which the lesion is located

in the superior cerebellar peduncle and one finds paralysis first, followed by ataxia.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

BRUSHFIELD'S SPOTS

Synonym: *Mongolian Spots*.

Ocular Findings: Pigment spots at the iris collarette in mongoloids.

Seen most readily in lightly colored irides.

They are found in 85 to 90% of mongoloids and only in about 20% of normal individuals.

The spots run in a circle around the iris and are due to elevations caused by a localized area of increased density of the anterior border layer associated with an accumulation of pigment.

In the mongoloid, the spots are located closer to the pupillary margin and are both more numerous and more distinct than in normal individuals. Also, the hypoplasia or thinning of the peripheral iris is prominent in mongoloids and unusual in normal individuals.

Bibliography

Brushfield, T.: Mongolism. *Brit. J. Child. Dis.*, 21:241-258, 1924.

Donaldson, D. D.: The significance of spotting of the iris in mongoloids: Brushfield's spots. *A.M.A. Arch. Ophthal.*, 65:26-31, 1961.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

BÜCKLER'S ANNULAR DYSTROPHY

Ocular Findings: Annular grayish lesions of the cornea primarily affecting Bowman's membrane.

Begins in early childhood.

The small opacities gradually merge into geographic configurations.

The overlying epithelium gradually changes and becomes irregular, often with recurring and very painful ulcerations.

The sensitivity of the cornea is reduced or abolished.

Vision is greatly reduced.

Etiology: Familial. Dominant inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BUERGER'S DISEASE

Synonym: *Thrombo-angiitis Obliterans*.

Ocular Findings: The ocular manifestations are complications of the systemic disease.

Transient blindness due to vasospasm.

Optic atrophy due to obliteration of the vascular supply to the optic nerve.

Recurrent vitreous hemorrhages.

Associated Clinical Findings: Generalized inflammatory disease of the vascular coats of the arteries and veins.

Vascular thrombosis and gangrene in the extremities.

Pathology: Vasculitis can be seen pathologically in the vessels of the optic nerve and in the retinal vessels.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 7. C. V. Mosby, St. Louis, Mo., 1962.

Hogan, M. J., and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

BUMKE'S SIGN

Synonym: *Bumke's Pupil Sign in Catatonia*.

Ocular Findings: Absence of dilation of the pupil on painful stimulus.

It is well known that a sudden, severe change in the physiologic state, such as due to fear, pain, joy or other excitement, causes a dilation of the pupils. This dilation of the pupil can be elicited experimentally by a painful stimulus such as sticking a pin in the face.

Pathology: The psychosensory pain reflex is absent or abnormal in idiocy, general paresis and, especially, catatonia.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examinations*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

BURGER-GRÜTZ DISEASE

Synonym: *Essential Familial Hyperlipemia.*

Ocular Findings: Retinal lipemia. Milky appearance of the vessels of the retina on ophthalmoscopic examination.
May see recurrent keratitis with fatty exudates.

Associated Clinical Findings: Hepatosplenomegaly.
Cutaneous xanthomatosis.
Abdominal cramps and nausea are frequent.
More commonly seen in children.

Pathology: Due to an increase in the content of neutral fats in the serum which thus becomes milky in appearance.

Etiology: Familial inheritance.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

BURNETT'S SYNDROME

Ocular Findings: Band Keratitis.

Associated Clinical Findings: Nephrocalcinosis.
Stomach symptoms.
Alkalosis.
Azotemia.
Seen frequently in patients on ulcer treatment.

Pathology: Hypercalcemia. A disease of abnormal calcium metabolism.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche

Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

BUSACCA NODULE

Ocular Findings: Pigmented nodule on the iris surface near the periphery seen in granulomatous uveitis.

Pathology: The nodules consists chiefly of epithelioid cells.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

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CAFFEY-SILVERMAN SYNDROME

Synonyms: *Caffey's Disease, Silverman-Caffey Disease, Infantile Cortical Hyperostosis.*

Ocular Findings: Soft tissue swelling around the eyes.

Tenderness around the eyes.

Transient proptosis has been reported.

May be unilateral or bilateral.

May have conjunctivitis.

Associated Clinical Findings: Soft tissue swelling of the jaws due to periosteal inflammation. May mimic mumps.

Tenderness and swelling may occur over the mandible, skull, clavicles, and ribs.

Occurs in infants up to age five or six months, and may be apparent at birth.

Course is benign and self-limited.

Diagnosis is made by x-ray.

Pathology: X-rays reveal cortical hyperostosis of the mandible, skull, clavicles and ribs.

Severe anemia is commonly found accompanied by elevated sed-rate and elevated serum phosphatase.

Etiology: Familial. The soft tissue swelling around the eyes is felt to be

due to periosteal inflammation, possibly by extension from the mandibular area.

Bibliography

- Barba, W. P. and Freriks, D.: The familial occurrence of infantile cortical hyperostosis in utero. *J. Pediat.*, 42:141-150, 1953.
 Caffey, J. and Silverman, W. A.: Infantile cortical hyperostosis; preliminary report of a new syndrome. *Amer. J. Roentgen.*, 54:1-16, 1945.
 Caffey, J.: Infantile cortical hyperostosis; a review of the clinical and radiographic features. *Proc. Roy. Soc. Med.*, 50:347-354, 1957.
 Minton, L. R.: Ophthalmic manifestations of infantile cortical hyperostosis (Caffey-Silverman disease). Presented to Southern Med. Soc., November 1966.
Yearbook of Pediatrics. Charles C Thomas, Springfield, Ill., 1965.

CAPGRAS' SYNDROME

Synonyms: *Illusion of Doubles Syndrome, Phantom Double Syndrome.*

Ocular Findings: Patient is unable to recognize an individual in his presence.

Patient steadfastly asserts that the person confronting him is double. As a compensatory measure he postulates a counterpart, describing differences between the subject before him and the alleged double.

Etiology: A form of schizophrenia.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
 Todd, J.: The syndrome of Capgras. *Psychiat. Quart.*, 31:250-265, 1957.

CAZENAVE'S DISEASE

Synonym: *Pemphigus Foliaceus.*

Ocular Findings: Loss of eyebrows.

Madarosis.

Ectropion.

Conjunctivitis.

Infiltration, vessicles and pannus formation on the cornea.

Iritis.

Cataracts in later stages of the disease.

Associated Clinical Findings: Erythematous or bullous lesions over the body followed by a generalized exfoliation of the skin.

Etiology: Unknown though infectious or toxic origin have been suggested.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

CESTAN SYNDROME

Synonyms and Similar Conditions: *Raymond-Cestan Syndrome, Cestan-Chenais Syndrome.*

Ocular Findings: Ocular muscle palsy caused by sixth nerve damage.

Associated Clinical Findings: Contralateral pyramidal tract involvement.
Posterior longitudinal bundle involvement.

Inferior cerebellar peduncle involvement.

Consequent contralateral hemiplegia.

Anesthesia of the face and extremities.

Findings almost identical to those described by some authors under the eponym Cestan-Chenais.

Pathology: Felt to be due to a thrombosis of a branch of the Basilar artery in the pons.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

CESTAN-CHENAIS SYNDROME

Synonyms and Similar Conditions: *Cestan Syndrome (may be a variation of the same thing), Babinski-Nageotti, Avellis Syndrome.*

Ocular Findings: Combination of eye signs of Babinski-Nageotti Syndrome and Avellis Syndrome.

Babinski-Nageotti: Enophthalmos, ptosis, nystagmus and miosis with hemiparesis.

Avellis: Horner's plus pharyngolaryngeal or glossopharyngeal paralysis.

Ocular findings occur on the side of the lesion; the cerebellar hemiataxia and sensory disturbances on the contralateral side.

Pathologic Etiology: May be caused by a thrombosis of the vertebral artery in proximity to the posterior inferior cerebellar and anterior spinal branches. Also may be seen in cerebellar pontine angle tumors.

Bibliography

- Cestan, R. and Chenais, L.: Du myosis dans certaines lésions bulbaires en foyer (hémiplégie du type Avellis associée ou syndrome oculaire sympathique). *Gaz. d. Hôp.*, 76:1229-1233, 1903.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

CHAGAS' DISEASE

Synonym: *Trypanosomiasis Cruzi*.

Ocular Findings: Acute lid edema.
Edematous conjunctivitis followed by keratitis.
May be unilateral or bilateral.

Associated Clinical Findings: Systemic signs of trypanosomiasis.
The acute form of this disease occurs mainly in children and is often ushered in after about two weeks of fever by Romana's sign.
The disease has a grave prognosis in children and is usually fatal in adults.

Pathology: Serious meningitis and edema of the brain.

Etiology: Infestation with *trypanosoma cruzi*.

Bibliography

- Atias, A.: *EENT Digest*, 27:20, 1965. (Abs).
Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

CHARCOT-MARIE-TOOTH SYNDROME

Synonym: *Progressive Muscular Atrophy*.

Ocular Findings: Variable nystagmus in all directions.
Occasional optic atrophy.

Vision diminished.

Loss of light reflex and irregularity of the pupils.

Occasional pigmentary retinopathy.

Associated Clinical Findings: Atrophy of the muscles beginning with the small muscles of the hands and feet, progressing to involve all of the muscles of the extremities.

Fibrillar twitching of the extremities.

Onset usually from age five to twenty.

Pathology: Degeneration in the motor cells of the spinal gray matter, pyramidal tracts and posterior columns.

Etiology: Familial. Male sex linked.

Bibliography

Hoyt, W. F.: Charcot-Marie-Tooth disease with primary optic atrophy; report of a case. *A.M.A. Arch. Ophthalm.*, 64:925-928, 1960.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

CHARCOT-WILBRAND SYNDROME

Synonym: *Visual Agnosia Syndrome.*

Ocular Findings: Visual agnosia.

Loss of ability to revisualize images seen.

Pathology: Occlusion of a portion of the posterior cerebral artery.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

CHARCOT'S AMYOTROPHIC LATERAL SCLEROSIS

Ocular Findings: Fine nystagmus.

Optic atrophy may occur.

Associated Clinical Findings: Rapidly progressing atrophy of the upper and lower limbs.

Muscular fibrillations.
Occasional glossopharyngeal paralysis.

Etiology: Familial. Dominant inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

CHARCOT'S TRIAD

Ocular Findings: Nystagmus, usually fine fibrillar.

Associated Clinical Findings: Intention tremor.
Scanning speech.

Pathologic Significance: A sign of cerebellar involvement in late multiple sclerosis.

Etiology: Familial. Males more affected.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

CHARLIN'S SYNDROME

Synonym: *Nasal Nerve Syndrome*.

Ocular Findings: Mucopurulent conjunctivitis.
Anterior uveitis and iritis.
Keratitis.
Corneal ulcers.
Severe orbital pain.

Associated Clinical Findings: Rhinorrhea.
Severe pain of the ala nasi.

Pathologic Etiology: Neuritis of the nasal branch of the trigeminal nerve.

Bibliography

Solotinitzky, I. N.: The syndrome of the nasal nerve (Charlin's Syndrome-complex).
ACTA Ophthal., 14:388-396, 1936.

CHÉDIAK-HIGASHI SYNDROME

Ocular Findings: Partial albinism of the iris and retina.

Visual loss.

Photophobia.

Papilledema.

Associated Clinical Findings: Lymphomatous development with hepatosplenomegaly.

Constitutional stigmata.

Defective resistance to infection.

Usually lethal early.

Pathology: Typical cytoplasmic inclusion bodies are present in many of the leukocytes.

Edema of the optic nerve and infiltration of the optic nerve with cells resembling immature lymphocytes.

Etiology: Familial. Usually occurs in albinoid siblings born of consanguinous parents.

Autosomal recessive.

Bibliography

Donohue, W. L. and Bain, H. W.: Chédiak-Higashi syndrome; a lethal familial disease with anomalous inclusions in the leukocytes and constitutional stigmata: report of a case with necropsy. *Pediatrics*, 20:416-430, 1957.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Johnson, D. L.; Jacobson, L. W.; Toyama, R.; *et al.*: Histopathology of eyes in Chédiak-Higashi syndrome. *Arch. Ophthalmol.*, 75:84-88, 1966.

CHERMAK'S TEST

Synonym: *Chermak's Sign.*

Ocular Findings: Visual projection of after-images presented to each eye.

In the presence of normal retinal correspondence, the patient will image a vertical line of light presented to the right eye, and a horizontal line of light presented to the left eye, as being a cross image by both eyes.

Thus Chermak's sign is the eponymic designation of the after-image confirmation of retinal correspondence, and is based on Chermak's after-image test.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examinations*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

CHEVASSE'S INHIBITIONAL PALSY

Synonym: *Paresis of Chevasse*.

Ocular Findings: In paresis of the contralateral antagonist, with the paretic eye fixing, the paresis may appear to involve the antagonist because:

- (1) When the antagonist contracts, its direct antagonist relaxes (according to Sherrington's law).
- (2) The yoke muscle of the antagonist (the contralateral antagonist) relaxes to the same degree (according to Hering's Law).

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

CIVANTOS SYNDROME

Synonyms and Similar Conditions: *20 Trisomy Syndrome, see Cockayne's Syndrome*.

Ocular Findings: Pigmentary retinopathy.
Photosensitivity.

Associated Clinical Findings: Progeroid facies.
Skeletal defects.
Nanism.
Mental retardation.

Etiology: Familial hereditary.

Bibliography

Francois, J.: Chromosome abnormalities and ophthalmology. *J. Pediat. Ophthalm.*, 1:5-18, 1964.

CLAUDE BERNARD'S SYNDROME

Synonym: *Bernard's Syndrome.*

Ocular Findings: Signs opposite to Horner's.

Ipsilateral pupillary dilatation.

Widening of the palpebral fissure (caused mainly by upper lid contraction).

Relative exophthalmos.

Associated Clinical Findings: May have decreased skin temperature ipsilateral. May have hyperhydrosis of ipsilateral side of face.

Pathologic Etiology: Caused by an irritative lesion involving any portion of the central or peripheral pathway of the sympathetic chain, in the midbrain, medulla or cervical cord.

Bibliography

Haessler, F.: *Eye Signs in General Disease*. Charles C Thomas, Springfield, Ill., 1960.
Lyle, D.: *Neuro-ophthalmology*. Charles C Thomas, Springfield, Ill., 1954.

CLAUDE'S SYNDROME

Synonym: *Rubro-ocular Syndrome. Closely resembles Benedikt's Syndrome.*

Ocular Findings: Unilateral paralysis of the third nerve.

Unilateral paralysis of the fourth nerve.

Resultant ocular pareses.

Associated Clinical Findings: Contralateral hemianesthesia.

Occasional hemiataxia.

Signs closely resemble Benedikt's.

Pathologic Etiology: Paramedian mesencephalic lesion starting in the midbrain.

Often occlusion of the terminal branches of the paramedian arteries supplying the inferior portion of the nucleus ruber.

Bibliography

Claude, H.: *Rev. Neurol.*, 1:1311, 1912.

COAT'S DISEASE

Synonym: *Retinitis Exudativa*.

Ocular Findings: Retinal telangiectasis (malformation of arterioles and venules).

Exudative retinitis.

Retinal hemorrhages.

Retinitis proliferans with iritis and complicated by glaucoma.

Secondary bullous detachment of the retina, usually unilateral.

Most occur in males and most in children.

Pathology: Fusiform dilation of the retinal veins, mainly in the periphery.

Exudative reaction gives the appearance of a yellowish white exudate mainly behind the vessels. The exudate may project several diopters and has been found to contain deposits of cholesterol.

The detachment usually begins in the lower portion of the fundus.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Howard, G.: Differential diagnosis of retinoblastoma. *Amer. J. Ophthal.*, 60:610-616, 1965.

Morales, A. G.: Coat's disease. *Amer. J. Ophthal.*, 60:855-864, 1965.

COAT'S WHITE RING

Synonym: *White Ring of the Cornea*.

Ocular Findings: A grayish-white ring seen in Bowman's membrane, usually small in diameter.

Usually appears just below the epithelium.

Can usually be related to trauma to the cornea.

Never interferes with vision.

Pathology: Lipoid and calcium salts in Bowman's membrane.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

COCKAYNE'S SYNDROME

Synonym: *20 Trisomy Syndrome.*

Ocular Findings: Pigmentary retinal degeneration.

Optic atrophy.

Sunken eyes, due to loss of orbital fat.

Cataracts.

Associated Clinical Findings: Dwarfism.

Mental retardation.

Dysacucia.

Nanism.

Skeletal defects such as kyphosis and ankylosis.

Early senile appearance.

Sensitivity to sunlight (photodermatitis).

Clinical onset at about age two.

Pathology: X-rays may show marbling of the epiphyses in some of the digits.

Etiology: Familial. Autosomal recessive.

Bibliography

Cockayne, E. A.: Dwarfism with retinal atrophy and deafness. *Arch. Dis. Child.*, 11:1-8, 1936.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Francois, J.: Chromosome abnormalities and ophthalmology. *J. Pediat. Ophthalm.*, 1:5-18, 1964.

Paddison, R. M.; Moossy, J.; Derbes, V. J.; *et al.*: Cockayne's syndrome. A report of five new cases with biochemical chromosomal dermatologic, genetic and neuropathologic observations. *Derm. Trop.*, 2:195-203, 1963.

COGAN'S MICROCYSTIC DYSTROPHY

Synonym: *Maplike Dystrophy of the Cornea.*

Ocular Findings: Non-staining benign epithelial dystrophy of the cornea.

Lesions appear as grayish white discreet or confluent round dot-like opacities measuring 0.1 to 0.5 mm in diameter located in the superficial cornea.

Usually appear with a map-like configuration.
More apparent in the central cornea but involves the entire cornea.
Causes slight reduction of vision.
Detectable only by slit lamp.

Pathology: Epithelium is loosely applied to Bowman's membrane because of an anomalous basement.
Contains discreet intraepithelial cysts containing pyknotic nuclei and cytoplasmic debris.

Etiology: Unknown. Usually appears after the fourth decade. Women primarily involved.

Bibliography

Cogan, D. G.; Donaldson, D. D.; Kuwabara, T.; *et al.*: Microcystic dystrophy of the corneal epithelium. Trans. Amer. Ophthal. Soc., 62:213-225, 1964.
Guerry, D.: Observations on Cogan's microcystic dystrophy of the corneal epithelium. Amer. J. Ophthal., 62:65-73, 1966.

COGAN'S SIGN

Ocular Findings: Engorgement of the veins over the lateral recti.
This is highly characteristic of the congestive type of dysthyroid exophthalmos.
The vessels appear as large tortuous sinusoids beginning and ending in the muscle or tendon.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

COGAN'S SYNDROME

Synonym: *Non-syphilitic Interstitial Keratitis.*

Ocular Findings: Patchy interstitial keratitis, later becomes vascularized.
Impaired vision.
Blepharospasm.
Lacrimation.
Conjunctival hyperemia.

Associated Clinical Findings: Abrupt onset is characteristic.
Deafness, and/or tinnitus.

Vertigo; vestibulo-auditory symptoms similar to Ménière's syndrome.
Nausea and vomiting with acute onset.
Usually affects young adults.

Pathologic Etiology: Unknown. May be related to periarteritis nodosa or vaccinia sensitivity. Non-syphilitic.

Bibliography

Cogan, D. G.: Non-syphilitic interstitial keratitis with vestibuloauditory symptoms; report of four additional cases. *Arch. Ophthalm.*, 42:42-49, 1949.

COGAN'S SYNDROME

Synonym: *Congenital Oculomotor Apraxia Syndrome.*

Ocular Findings: The eye cannot be turned voluntarily to one side and the patient attempts to compensate by also turning the head toward the object whereupon the vestibular reflex causes the eyes to deviate even further from the object.

Further turning of the head results in overshooting the object, but by means of compensatory, jerky movements, fixation is finally accomplished in about one second.

Seen more commonly in young children.

Etiology: Unknown. The prognosis is good.

Bibliography

Cogan, D. G.: Type of congenital ocular motor apraxia presenting jerky head movements. *Amer. J. Ophthalm.*, 36:433-441, 1953.

COLLIER'S SIGN

Ocular Findings: Retraction of the eyelids, mild (Collier's Sign).

Accompanying look of pseudo-cheerfulness and an alert expression.

Associated Clinical Findings: No evidence of hyperthyroidism.

Pathologic Etiology: Diencephalic neoplasm.

Bibliography

Keitel, H. G.; Girone, J. A. C.; Soentgen, M. L.; *et al.*: Peculiar versus diagnostic facies in pediatrics. *Lippincott's Med. Sci.*, 17:47-58, 1966.

COLLINS-FRANCESCHETTI-ZWAHLEN SYNDROME

Synonym: *Mandibulofacial Dysostosis.*

Ocular Findings: Palpebral fissures slope downward, outward.
Lower lids may have a colobomatous notch.
Stabismus is frequent.

Associated Clinical Findings: Face is narrowed.
Ears are deformed.

Etiology: Familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

CONRADI'S SYNDROME

Ocular Findings: Bilateral total cataracts.
Cataracts develop in infancy.

Associated Clinical Findings: Congenital stippled epiphyses.
Short legs.
Patient appears as a short limbed dwarf with immobile joints and flexion contractures leading to immobility of the extremities.

Pathology: X-rays show spotty calcification of the epiphyses.

Etiology: Unknown. Probably familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

COPPOCK CATARACT

Synonym: *Nuclear Cataract.*

Ocular Findings: This is a grayish white diffuse central lenticular opacity visible best with lateral illumination.
With the biomicroscope one can see that it is composed of many

small white dots dispersed throughout the embryonic nucleus.
Vision may be reduced.

This has also been called a discoid cataract, or central cataract.

Etiology: Felt to be a dominantly inherited characteristic.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

COWEN'S SIGN

Ocular Findings: Jerky pupillary reaction to light, in hyperthyroidism.
Felt to be a jerky form of the consensual pupillary reflex.

Associated Clinical Findings: Graves' disease.

Tachycardia, trembling, increased basal metabolism, and other signs
of hyperthyroidism.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed.
Grune and Stratton, New York, N.Y., 1961.

CREIX-LÉVY SYNDROME

Synonym: *Ophthalmosomato-hygrosis*.

Similar Condition: *Barré-Lieou Syndrome*.

Ocular Findings: Excessive lacrimation.
Eye ache.

Associated Clinical Findings: Excessive rhinorrhea.

Vasomotor changes in the skin.

Pain in the cervical area.

Excessive salivation.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche
Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45,
Feb., 1966.

CREUTZFELD-JACOB DISEASE

Synonym: *Cortico-striato-spinal Degeneration.*

Ocular Findings: Anisocoria.

Nystagmus.

Extraocular paralyses may develop.

Associated Clinical Findings: Weakness in the extremities.

Unsteadiness of gait.

Choreoathetosis occurs later.

Progresses to mental deterioration, aphasia, epileptiform seizures, emaciation and death.

Pathology: Widespread degenerative changes involving the cerebral cortex in its deeper layers and the basal ganglia.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

CROUZON'S DISEASE

Synonym: *Dysostosis Craniofacialis.*

Ocular Findings: Exophthalmos.

Obliquity of the palpebral fissures with the outer canthus slanting downward.

Nystagmus.

Divergent strabismus.

Optic atrophy and visual loss.

Visual fields show upper field defects due to pressure on the optic nerve at the foramen.

Sclera often appears bluish.

Associated Clinical Findings: Maxillary atrophy.

Prognathism.

Enlarged nasal bones and bony abnormalities in the region of the superior longitudinal sinus and bregma.

Frequent headaches.

Subnormal mentality.

Occasional hearing loss.

Etiology: Congenital.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

Crouzon, O.: Dysostose cranio-faciale héréditaire. Bull. et mém. Soc. Méd. des Hôp. Paris, 33:545-555, 1912.

CROWE'S SIGN

Synonym: *Cavernous Sinus Thrombosis Sign.*

Ocular Findings: Retinal venous engorgement with jugular pressure opposite a thrombosis.

Cavernous sinus thrombosis confined to one side in its early stages does not produce venous engorgement, but bilateral obstruction to venous outflow always results in venous engorgement.

Consequently, pressure on the internal jugular vein on the side opposite the suspected thrombosis results in bilateral retinal venous engorgement.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

CUSHING'S SYNDROME

Synonyms: *Angle Tumor Syndrome, Cerebellopontine Angle Syndrome.*

Ocular Findings: Paresis of the external rectus muscle (usually unilateral).

Mixed nystagmus.

Decreased corneal reflex.

Associated Clinical Findings: Paresis of the orbicularis muscle (seventh nerve paralysis)

Deafness.

Tinnitus.

Facial hyperesthesia.

Hoarseness.

Dysphagia.

Pathologic Etiology: Usually caused by a tumor of the cerebellopontine angle.

Bibliography

Brown, J. R.: Localizing cerebellar syndromes. JAMA, 141:518-521, 1949.

CUSHING'S SYNDROME

Synonym: *Cushing's Chiasmal Syndrome.*

Ocular Findings: Early primary optic atrophy.

Bitemporal field defects.

Occasional papilledema.

Pathology: X-ray shows normal sella turcica.

Internal hydrocephalus may occur from blocked foramen of Monroe with consequent papilledema.

Etiology: May be caused by supracellar cysts, gliomas, meningioma, or aneurysm.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Cushing, H.: Chiasmal syndrome of primary optic atrophy and bitemporal field defects in adults with normal sella turcica. A.M.A. Arch. Ophthal., 3:505, 1930.

D

DALEN-FUCH'S SPOT

Synonym: *Dalen-Fuch's Nodule.*

Ocular Findings: Small grayish-brown nodules appearing in the retina periphery in cases of sympathetic ophthalmitis.

Produce small round areas of depigmentation on recovery (the Dalen-Fuch's spot).

Pathology: The nodules are primarily in the pigment epithelium and consist chiefly of epithelioid cells.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

DALRYMPLE'S SIGN

Ocular Findings: Abnormal wideness of the palpebral fissure due to upper lid retraction.

Gives the patient a staring look.

Becomes more pronounced when the patient is consciously fixating an object.

Occurs in more than one-half of patients with thyrotoxicosis and is usually the earliest sign of Graves' disease.

Associated Clinical Findings: Graves' disease.

Tachycardia, trembling, increased basal metabolism and other systemic signs of hyperthyroidism.

Bibliography

Haessler, F. H.: *Eye Signs in General Disease*. Charles C Thomas, Springfield, Ill., 1960.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

DANDY-WALKER SYNDROME

Synonym: *Syndrome of Atresia of the Foramen of Magendie.*

Ocular Findings: Ptosis.

Sixth nerve paralysis.

Papilledema.

Associated Clinical Findings: Usually seen in infants.

Hydrocephalus.

Prognosis poor.

Pathology: Malformation and stenosis of the foramina of Luschka and Magendie with dilation of the fourth ventricle and anomalies of the rostral portion of the vermis.

Bibliography

Gardner, W. J.; Abdullah, A. F.; and McCormack, L. J.: The varying expressions of embryonal atresia of the fourth ventricle in adults: Arnold-Dandy-Walker syndrome, arachnoid cyst of the cerebellum, and syringomyelia. *J. Neurosurg.*, 14:591-605, 1957.

Keet, P. C. and Berman, M. C.: The Dandy-Walker syndrome, a report of three cases. *So. Afr. Med. J.*, 35:6-11, 1961.

DARIER'S DISEASE

Ocular Findings: Follicular keratitis.

May have lacrimal insufficiency.

Photophobia.

May have absence of Bowman's membrane.

May be complicated by pannus formation.

Associated Clinical Findings: Cutaneous telangiectasia.

Myxedema.

Crusting, ulcerating papules of the skin.

Etiology: Vitamin A deficiency.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

DEJEAN'S SYNDROME

Synonym: *Orbital Floor Syndrome.*

Ocular Findings: Exophthalmos.

Diplopia.

Associated Clinical Findings: Pain in the superior maxillary region.

Numbness in the area of the first and second branches of the trigeminal nerve.

Pathologic Etiology: Space-occupying lesion involving the floor of the orbit. Extension of the lesion into the cranial cavity may occur.

Bibliography

Dejean, C.: Le syndrome du plancher de l'orbite. Bull, et mém. Soc. Franc. d'ophthal., 48:473-485, 1935.

DÉJERINE-KLUMPKE SYNDROME

Synonyms: *Lower Radicular Syndrome, Klumpke's Syndrome.*

Ocular Findings: Enophthalmos.

Ptosis.

Miosis.

Associated Clinical Findings: Paralysis and atrophy of the small muscles of the forearm and hand.

Pathology: Lesion involving the inferior roots of the brachial plexus with nerves derived from the eighth cervical and first thoracic roots.

Etiology: May be caused by birth trauma, Potts disease, Pancoast's tumor, hemorrhage, myelitis, etc.

Bibliography

Dejerine: *Rev. Med. (Paris)*, 5:591, 1885.

Haessler, F. H.: *Eye Signs in General Disease*. Charles C Thomas, Springfield, Ill., 1960.

DÉJERINE-SOTTAS DISEASE

Synonym: *Hypertrophic Neuritis*.

Ocular Findings: Pupillary disturbances such as myosis, anisocoria or pupillotonia.

Occasionally optic atrophy.

Associated Clinical Findings: Polyneuritis.

Paresis involving the extremities.

Muscular atrophy.

Sensory disturbances of the peripheral type.

May have tabetiform symptoms with lightning pains and visceral crises.

Etiology: Hereditary. Regular dominant transmission.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

DE LANGE SYNDROME

Ocular Findings: Eyebrows hypertrichotic and joined in the midline.

Eyelashes long.

Synorphys.

Hypertelorism or telecanthus,

Ptosis.

High myopia.

Antimongoloid slant of the palpebral fissures.

Associated Clinical Findings: Mental retardation.

Growth retardation or dwarfism.

Skeletal abnormalities such as syndactyly.

Hirsutism of the forehead and back.

Low hairline.

Feeble, raucous cry in infancy.

Nose small and upturned.

Distance between nose and upper lip greater than usual.

Lips thin and down curved.

Etiology: Unknown. Congenital.

Bibliography

Hart, Z. H., *et al.*: The de Lange syndrome. Amer. J. Dis. Child., 109:325-332, 1965.

Nicholson, D. H. and Goldberg, M. F.: Ocular abnormalities in the de Lange syndrome. Arch. Ophthal., 76:214-220, 1966.

DENNIE-MARFAN SYNDROME

Synonym: *Congenital Syphilitic-paralysis Syndrome.*

Ocular Findings: May have nystagmus.

Frequently associated with cataracts.

Associated Clinical Findings: Most frequent in children with congenital syphilis.

Spastic paralysis.

Mental retardation.

Frequent occurrence of epilepsy.

Etiology: Congenital syphilis.

Bibliography

Dennie, C. C.: Partial paralysis of the lower extremities in children, accompanied by backward mental development. Amer. J. Syph., 13:157-163, 1929.

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

DEVIC'S DISEASE

Synonym: *Neuromyelitis Optica*.

Ocular Findings: Bilateral acute optic neuritis.

Rapid progressive loss of vision.

Visual recovery in fifty per cent.

Associated Clinical Findings: Most frequent between age 20 to 30.

Pain in the extremities.

Numbness and weakness of the extremities.

Progresses to paraplegia.

Pathology: Demyelination of the nerves with destruction of the axis cylinders, microglial proliferation, astrocytosis, and perivascular lymphocytosis.

Affects the optic tract and the spinal cord.

Etiology: Unknown. Considered by some to be a form of acute multiple sclerosis.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Orteza, J.: A case of ependymoma simulating Devic's syndrome; a clinicopathological case report. A.M.A. Arch. Ophthalm., 64:940-945, 1960.

Stansbury, F. C.: Neuromyelitis optica (Devic's disease); presentation of five cases, with pathologic study, and review of literature. Arch. Ophthalm., 42:292-335, 1949.

DIMITRI'S DISEASE

Synonyms: *Sturge-Weber-Kalisher-Dimitri Syndrome, Encephalo-trigeminal Angiomatosis*.

Ocular Findings: Angiomas of the choroid.

May show hydrophthalmos.

May be complicated by glaucoma.

Port-wine hemangioma of the eyelid.

Associated Clinical Findings: Usually unilateral nevus flammeus (hemangioma of the skin), distributed along branches of the fifth nerve.

Mental retardation.
May have convulsions.

Pathology: X-rays show laminated calcification in affected areas of the skull.

Etiology: Felt to be a partial trisomy of a D chromosome. Hereditary.
Simple recessive.

Bibliography

Datelines in Ophthal., Vol. 2, No. 1, 1965.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

DIMMER'S NUMMULAR KERATITIS

Synonym: *Keratitis Nummularis*.

Ocular Findings: Slowly-developing benign keratitis without accompanying conjunctivitis.

Occurs sporadically and usually unilaterally.

Characterized by disc-shaped infiltrates in the superficial corneal stroma.

Incidence is seasonal (mainly autumn).

Associated with irritation, photophobia, and lacrimation.

Vision is diminished.

May be progressive for years.

Prognosis is good.

Etiology: Probably viral. May follow trivial trauma.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

DOWN'S SYNDROME

Synonyms: *Mongolism*, *"21 Trisomy" Syndrome*, *"D Trisomy" Syndrome*.

Ocular Findings: Hypertelorism.
Small and oblique eyelid fissures.

Epicanthus.

Yellow spots on the iris (Brushfield).

Lens opacities in fifty per cent.

High myopia in thirty per cent.

Associated Clinical Findings: Mental retardation.

Skeletal abnormalities and retarded growth.

Excessive salivation and drooling.

Hyperflexion of joints.

Etiology: Familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Ginsberg, J. and Perrin, E. V. D.: Ocular manifestations of 13-15 trisomy. *Arch. Ophthalm.*, 74:487-495, 1965.

DOYNE'S CHOROIDITIS

Synonym: *Doyme's honeycomb degeneration.*

Ocular Findings: Degeneration of the retina.

Characterized by the presence in the papillomacular region of rounded grayish-white spots, very close together, forming a mosaic and presenting a "honeycomb" appearance.

As the condition progresses, central vision is lost.

Pathology: Colloid deposits between the macula and the optic disc.

Etiology: Familial. More frequent in older women.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

DOYNE'S GUTTATE IRITIS

Ocular Findings: Keratic precipitates on the iris.

Appear as small gray masses on the anterior surface of the iris.

Rarely may appear on the anterior capsule of the lens.
Frequently accompanied by faint anterior chamber flare.

Pathologic Etiology: Evidence of low-grade nonspecific anterior uveitis.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

DRESBACH'S SYNDROME

Synonym's: *Herrick's Syndrome, Sickle Cell Disease.*

Ocular Findings: Vitreous hemorrhages.

Complicated cataract.

Secondary glaucoma.

Exudative and hemorrhagic retinopathy.

May see angioid streaks of the retina.

Associated Clinical Findings: Sickle cell anemia.

Moderate jaundice.

May have episodes of severe abdominal pain simulating a surgical condition of the abdomen.

Hemiplegia, cranial nerve palsies and other neurologic disturbances may result from cerebral thromboses.

Pathology: Sickled erythrocytes. Hyperplastic bone marrow.

Etiology: Inherited. Dominant transmission. Occurs almost exclusively in the Negro race.

Bibliography

Goodman, G.; Von Sallmann, L.; and Holland, M. G.: Ocular manifestation of sickle cell disease. A.M.A. Arch. Ophthal., 58:655-682, 1957.

DUANE'S SYNDROME

Synonyms: *Duane's Retraction Syndrome, Stilling's Syndrome, Turk-Still-ing Syndrome.*

Ocular Findings: Deficient horizontal recti motility with limitation of abduction with retraction of the globe on attempted adduction of the affected eye.

Adduction of the eye is associated with narrowing of the palpebral fissure.

Abduction of attempted abduction is accompanied by widening of the palpebral fissure.

Eye may be esotropic in the primary position.

Forced duction test shows restriction of movement in abduction or adduction or both.

Associated Clinical Findings: Malformations of the face and ears.

May be associated with Klippel-Feil syndrome.

Pathology: Thought to be a degenerative fibrosis of the lateral rectus muscle.

Etiology: Cause uncertain. Female sex linkage.

Bibliography

Alexander, C. M.: Bilateral Duane's retraction syndrome. *Amer. J. Ophthalm.*, 60:907-910, 1965.

Blodi, F.; Allen, W. Van; and Yarbrough, J. C.: Duane's syndrome. A brain stem lesion. *Arch. Ophthalm.*, 72:171-177, 1964.

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Costa Fernandes, Ruy: Duane's syndrome. *Amer. J. Ophthalm.*, 36:146, 1953.

DUHRING'S DISEASE

Synonym: *Dermatitis herpetiformis*.

Ocular Findings: Pseudomembranous conjunctivitis, resembling that seen in erythema multiforme but less frequently present.

When the conjunctiva is involved it is usually severe and produces pseudomembranes, cicatrization, and shrinkage of the cul-de-sac.

Associated Clinical Findings: Patchy erythematous eruptions of the skin.

May have headaches, malaise, joint and muscle pains, and fever.

Etiology: Cause not certain. Felt to be infective.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

E

EALES DISEASE

Ocular Findings: Recurrent massive retinal and vitreous hemorrhages.
Usually involves the left eye first.
About one half progress to blindness.
May be complicated by retinitis proliferans.
May be complicated by retinal detachment.
Occurs mostly in young males (average age about 23).

Pathology: Periphlebitis with vascular sheathing and telangiectasis.

Etiology: Unknown, but frequently found in association with tuberculosis. However the bacteriologic proof of its being tuberculous lacks confirmation.

Bibliography

- Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.
Jain, B. S. and Srivastava, K. N.: Eales disease followed by solitary choroidal tubercle. *Eye Ear Nose Throat Monthly*, 45:70-71, 1966.

ECKMAN-LOBSTEIN SYNDROME

Synonyms and Similar Conditions: *Osteogenesis Imperfecta, Lobstein's Disease, Van Der Hoeve's Syndrome.*

Ocular Findings: Blue sclera, felt to be due to thinning and partial transparency.
May have keratoconus or megacornea.
May have cataracts.

Associated Clinical Findings: Deafness appearing in early adulthood.
Bony fragility resulting in multiple spontaneous fractures.

Etiology: Familial.

Bibliography

- Arthur, L. J. H.: Some hereditary syndromes that include deafness, *Develop. Med. Child. Neurol.*, 7:395-409, 1965.

Kearns, T. P.: Neuro-ophthalmology. Annual Review, Arch. Ophthal., 76:729-755, 1966.

EDWARDS' SYNDROME

Synonym: "18 Trisomy Syndrome," "E Trisomy Syndrome."

Ocular Findings: Corneal opacities.
Ptosis.

Associated Clinical Findings: Mental retardation.
Low-set ears.
Hyperflexion of fingers.
Constitutional weaknesses.

Pathologic Etiology: Familial. Patients have a karyotype of 47 chromosomes with a trisomy for chromosome number 18.

Bibliography

Ginsberg, J. and Perrin, E. V. D.: Ocular manifestations of 13-15 trisomy. Report of case with clinical cytogenetic and pathologic findings. Arch. Ophthal., 74:487-495, 1965.

EHLERS-DANLOS SYNDROME

Synonyms: *Fibrodysplasia Elastica Cutis, Hyperelastica, Rubber-skin Disease.*

Ocular Findings: Blue sclera due to thinning of the ocular tissues.
Angioid streaks of the retina.
Depigmentation of the choroid.
Retinal hemorrhages.
May be complicated by retinitis proliferans.
May have hypotony of the extraocular muscles and strabismus.

Associated Clinical Findings: Hyperelasticity and fragility of the skin.
Hyperextensibility of the joints.
Muscular hypotony.
May have hypercalcemia.

Pathology: Degeneration of elastic mesenchymal tissue.

Etiology: Familial. Transmitted as an irregular autosomal dominant condition.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.
- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
- Johnson, S. A. M. and Falls, H. F.: Ehlers-Danlos syndrome; clinical and genetic study. *Derm. and Syph.*, 60:82-105, 1949.
- Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

EHRlich'S LINE

Synonym: *Ehrlich-Turk Line*.

Ocular Findings: A relatively thin vertical line of pigmented keratic precipitates deposited on the endothelium of the lower cornea.
It is a sign of low grade anterior uveitis.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

ELSchnig BODIES

Synonym: *Elschnig Pearls*.

Ocular Findings: A type of after-cataract due to epithelial cell proliferation with formation of clear grape-like clusters in the area of the lens periphery.

These epithelial pearls form following extracapsular cataract extractions in which remnants of the posterior capsule remain.

Bibliography:

- Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.
- Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

ELSchnig SPOTS

Synonym: *Elschnig dots*.

Ocular Findings: Black isolated flecks of pigment associated with bright yellow or red halo from choroiditis.

Seen in cases of advanced nephritis.

Pathology: Hypertrophic spots overlying sclerosed choroidal vessels.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 7. C. V. Mosby, St. Louis, Mo., 1962.

ELSCHNIG'S SYNDROME

Ocular Findings: Ectropion of the lower eyelids.
Especially long palpebral fissures.
Lateral canthal angles are displaced downward.

Associated Clinical Findings: Cleft hard palate.
Hare lip.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

ENROTH'S SIGN

Ocular Findings: Puffy edema of the lids.
Seen especially in more severe cases of thyrotropic disease.

Associated Clinical Findings: Graves' disease.
Tachycardia, trembling, increased basal metabolism, and other signs of hyperthyroidism.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

ERB-GOLDFLAM'S DISEASE

Synonyms: *Myasthenia Gravis*, *Asthenic Bulbar Paralysis*.

Ocular Findings: Diplopia.
Ptosis.
Strabismus and occasionally complete ophthalmoplegia externa.

Associated Clinical Findings: Excessive fatiguability of striated muscles, first manifesting itself in the muscles innervated by the bulbar nuclei.

Dysphagia.

Dysphonia.

General muscular exhaustion after slight activity.

Mortality eighty per cent within two years of onset.

Etiology: Unknown. Females affected two to one over males.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Grob, D.: Course and management of myasthenia gravis. *JAMA*, 153:529-532, 1953.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

ESPILDORA-LUQUE SYNDROME

Synonym: *Ophthalmic Sylvian Syndrome*.

Ocular Findings: Unilateral blindness.

Associated Clinical Findings: Temporary hemiplegia on contralateral side of the amaurosis, caused by a reflex spasm of the middle cerebral artery.

Pathologic Etiology: Embolus of the ophthalmic artery.

Bibliography

Espildora-Luque, C.: Ophthalmic Sylvian syndrome. *Amer. J. Ophthal.*, 18:402, 1935.

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

F

FABRY'S DISEASE

Synonym: *Glycolipid lipidosis*.

Ocular Findings: Puffy eyelids.

Periorbital edema.

Corneal epithelial hazy opacities.

Nuclear cataracts in about fifty per cent.
Vision not reduced.
Irregular conjunctival vessel dilations.

Associated Clinical Findings: Burning sensations or even severe pain in the extremities.
Fever after brief exercise or in hot weather.
Peculiar dark purple skin eruption with a "bathing-suit" distribution (genitalia, thighs, buttocks and navel).
Diffuse malaise.
Renal disease from which affected males die prematurely.

Pathology: Glycolipid is deposited in various tissues throughout the body.

Etiology: Sex linked recessive. Males primarily affected.

Bibliography

Spaeth, G. and Frost, P.: Fabry's disease. Arch. Ophthal., 74:760-768, 1965.
Spaeth, G.: *Datelines in Ophthal.*, 2:7, 1965.

FANCONI'S DISEASE

Synonym: *Fanconi's aplastic anemia.*

Ocular Findings: Convergent strabismus.
May have a band keratopathy.

Associated Clinical Findings: Spots of brown pigmentation on the skin.
Retarded physical development with skeletal anomalies.
Genital hypoplasia.
Tendon hyperreflexia.
Microcephalia.

Pathology: This is a grave hyperchromic aplastic anemia which appears in children between five and seven years of age.

Etiology: Familial. Simple recessive inheritance.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FANCONI-TÜRLER SYNDROME

Ocular Findings: Congenital ocular dysmetria (uncoordinated eye movements).

Nystagmus of the supranuclear type.

Associated Clinical Findings: Cerebellar ataxia.

Seen in early infancy.

Etiology: Familial. Possibly due to consanguineous marriage.

Bibliography

Fanconi, G. and Türler, U.: Kongenitale kleinhirnatrophie mit supramucleären störungen der motilität der augenmuskeln. Helvet. Paediat. ACTA., 6:475-483, 1951.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FAVRE'S DISEASE

Synonym: *Lymphopathia, Granuloma Venerium.*

Ocular Findings: Edema and infiltration of the eyelids.

Occasional keratitis.

Pannus may be present.

Associated Clinical Findings: Inguinal buboes.

Frequent in Negroes, rare in whites.

Etiology: Specific virus. Frei skin test positive.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

FEER'S SYNDROME

Synonyms: *Feer's Vegetative Neurosis, Swift's Disease, Infantile Acro-dynia.*

Ocular Findings: Photophobia with or without conjunctivitis.

Proptosis is present in about fifty per cent of cases.

Associated Clinical Findings: Seen in older infants.

Erythema, eruptions and hyperkeratosis of the skin.

Cyanosis of the extremities.

Weakness, irritability and hyperesthesia.

Profuse sweating.

Chronic but prognosis is good.

Etiology: Uncertain but felt to be infectious.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

FEHR'S SPOTTED DYSTROPHY

Synonym: *Groenouw's Dystrophy Type Two.*

Ocular Findings: Progressive dystrophy of the cornea beginning in the first decade with dullness and irregularity of the corneal epithelium caused by small indistinct grayish opacities in the central area of the cornea superficially.

In the second and third decades, the opacities appear in the deeper layers over the entire cornea.

Later, the entire cornea shows a diffuse cloudiness due to irregular masses of opacities in all layers of the cornea.

Corneal sensitivity is reduced.

Recurring erosions of the cornea may occur.

Etiology: Familial. Autosomal recessive inheritance.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FELTY'S SYNDROME

Similar Condition: *See Still's Disease.*

Ocular Findings: Kerato-conjunctivitis sicca.

Episcleritis.

Hypopyon ulcer.

Occasionally scleromalacia perforans.

Associated Clinical Findings: Chronic polyarthritis.

Splenomegaly.

Leucopenia.

Skin pigmentations are reported.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

FIESSINGER-LEROY SYNDROME

Synonyms: *Reiter's Syndrome, Conjunctivo-urethro-synovial Syndrome.*

Ocular Findings: Sterile purulent conjunctivitis.

Occasional iritis.

Occasional keratitis.

Associated Clinical Findings: Arthritis with pain, swelling, heat, and effusion.

Urethritis.

Peripheral lymphadenopathy.

Genital and oral ulcerations.

Occasional skin erythema.

Anorexia.

Prognosis good with treatment.

Etiology: Unknown. Affects young adult males primarily.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Foxworthy, D. T.: Adrenocorticotropin and cortisone in treatment of severe Reiter's syndrome. *Ann. Int. Med.*, 44:52-62, 1956.

FIESSINGER-RENDU SYNDROME

Synonym: *Ectodermosis Erosiva Pluriorificialis.*

Similar Conditions: *Stevens-Johnson disease, Fuchs' mucocutaneous-*

ocular syndrome, felt to be a form of Erythema multiforme. See Hebra.

Ocular Findings: Purulent or pseudomembranous conjunctivitis.

May have keratitis.

Xerosis may be present.

May be complicated by symblepharon.

Associated Clinical Findings: Papulo-vesicular or hemorrhagic eruptions of the extremities with multiform lesions.

Stomatitis.

May have urethritis, balanitis or vulvovaginitis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl., Augenheilk., 148:1-45, Feb., 1966.

FISCHER-KHUNT SPOT

Synonym: *Senile Hyaline Plaque.*

Ocular Findings: Bluish transparency of the sclera anterior to the insertions of the horizontal recti muscles.

Pathologic Etiology: Represents a thinning and hyalinization of the sclera.

No associated pathology.

FISHER'S SYNDROME

Synonym: *Ophthalmoplegia-ataxia-areflexia Syndrome.*

Ocular Findings: Sudden progressive complete external ophthalmoplegia, usually bilateral.

May have partial internal ophthalmoplegia.

Associated Clinical Findings: Ataxia.

Loss of deep reflexes.

Usually follows an upper respiratory infection.

Findings similar to Guillain-Barre syndrome.

Prognosis is good. Complete recovery is the rule.

Pathologic Etiology: Due to hypersensitivity to a neurotropic viral infection.

Bibliography

Fisher, M.: An unusual variant of acute idiopathic polyneuritis (syndrome of ophthalmoplegia, ataxia and areflexia). *New Eng. J. Med.*, 255:57-65, 1956.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FLEISCHER'S DYSTROPHY

Synonyms: *Cornea verticillata, Fleischer's granular dystrophy type two.*

Similar Conditions: *Felt to be the same as Groenouw's type one.*

Ocular Findings: Granular hyaline-like material deposited between the lamellae of the corneal stroma.

Characterized by the appearance in Bowman's membrane of innumerable brownish points arranged in curved lines converging toward the center of the cornea and giving the impression of a whirlpool.

Etiology: Familial. Dominant inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FLEISCHER RING

Ocular Findings: A brownish-to-yellow pigment ring seen in Bowman's membrane surrounding the base of a keratoconus cone.

Usually centered inferior and nasal to the central cornea.

Pathology: Pigment is felt to be hematin.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

FLOUREN'S SIGN

Synonym: *Flouren's Law*.

Ocular Findings: Nystagmus occurring in the plane of an affected semicircular canal.

Flouren's law states that "each semicircular canal gives rise to nystagmus in the plane of that canal."

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

FOIX SYNDROME

Synonyms: *Cavernous Sinus Syndrome*, *Hypophyseal-sphenoidal Syndrome*.

Ocular Findings: Proptosis.

Lid edema.

Paralysis of the third, fourth, fifth, and sixth nerves.

Chemosis of the globe.

Trigeminal neuralgia.

Optic atrophy.

Associated Clinical Findings: Occasional post-auricular edema.

Pathologic Etiology: Tumor, aneurysm, thrombosis, or inflammatory lesion involving the lateral cavernous sinus wall or the sphenoid bone.

Bibliography

Goldman, M. and Adams, R. D.: Fibrosarcoma of sphenoid bone, producing syndrome of lateral wall of cavernous sinus—case report. *J. Neuropath. Exp. Neurol.*, 5:155-159, 1946.

FÖLLING'S SYNDROME

Synonym: *Oligophrenia*.

Ocular Findings: Partial ocular albinism.

Photophobia.

May have cataracts.

May have corneal opacities.

Associated Clinical Findings: Mental retardation.

Partial general albinism.

Extrapyramidal motor symptoms.

Infantilism.

Pathology: Failure to metabolize phenylalanine so that this amino acid is excreted in the urine.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

FORSTER'S DIFFUSE CHOROIDITIS

Ocular Findings: Granulomatous posterior uveitis.

Posterior vitreous appears hazy.

May have grayish-yellow area of hemorrhage in the posterior pole.

Ring scotoma with macular sparing.

Vision diminished.

Pathology: Retinal periarteritis.

Etiology: Syphilitic.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

FORSTER-FUCHS' SPOT

Synonym: *Forster-Fuchs' Black Spot*.

Ocular Findings: An unusual amount of pigmentation in the region of the macula.

Associated with moderate to high myopia.

FOSTER-KENNEDY SYNDROME

Synonyms: *Gowers-Paton-Kennedy Syndrome, Basal-frontal Syndrome*.

Ocular Findings: Central scotoma, retrobulbar neuritis and optic atrophy on the homolateral side of lesion.

Papilledema in the contralateral eye.

Associated Clinical Findings: May have anosmia.

Pathology: Tumor, abscess of aneurysm in the base of the frontal lobe or sphenoidal area. Presses down on underlying structures. The prognosis depends on the causative lesion.

Bibliography

Masters, S.: Foster-Kennedy syndrome. *Amer. J. Ophthal.*, 36:983-985, 1953.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

FOVILLE'S SYNDROME

Similar Condition: *See Millard-Gubler Syndrome.*

Ocular Findings: Paralysis of conjugate movement to the side of a destructive lesion in the pons.

Homolateral sixth and eighth nerve palsy.

Deviation of the eyes away from the side of lesion.

During recovery gross nystagmus may be seen.

Associated Clinical Findings: Contralateral hemiplegia.

Peripheral facial palsy.

Pathologic Etiology: Interruption of the cortical fibers to the cranial nerve nuclei.

Clinical manifestations may be caused by pontine area tumor, hemorrhage, multiple sclerosis, tuberculosis, etc.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

FRANCESCHETTI SYNDROME

Synonym: *Mandibulofacial Syndrome.*

Ocular Findings: Underdeveloped orbital rim.

Ectropion.

Blepharophimosis.

Lid colobomas, especially of the lateral third of the lower lids.
Eyes appear to slope down laterally (antimongoloid).

Associated Clinical Findings: Hypoplasia of the facial bones.
High palate and abnormal dentition.
May have cleft palate.
Malformed ears.
Occasional hearing loss.

Etiology: Familial. Irregular dominant transmission.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.
Franceschetti, A. and Klein, D.: Mandibulo facial dysostosis; new hereditary syndrome. *ACTA. Ophthal.*, 27:143-224, 1949.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FRANCESCHETTI'S CORNEAL DYSTROPHY

Synonym: *Deep punctate dystrophy*.

Ocular Findings: Dystrophy of the deeper layers of the corneal stroma close to Descemet's membrane, consisting of very small whitish spots associated with thin filaments or rod-shaped forms.

Associated Clinical Findings: Only two cases reported. Both also had ichthiosis.

Etiology: Possibly familial.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

FRANCESCHETTI'S FLECKED RETINA SYNDROME

Synonym: *Fundus flavimaculatus*.

Ocular Findings: Yellow or yellow-white areas in the retina similar or identical to what is seen in some of the tapetoretinal degenera-

tions, but in which the diagnosis of a tapetoretinal degeneration is not borne out by electrophysiologic and psychophysical tests. Associated with macular degeneration but may retain full peripheral fields.

May have central or paracentral scotomas.

Peripheral lesions are of small, irregularly shaped atrophic areas deep in the retina in which the pigment epithelium has been lost, and in or around which pigment is clumped or scattered.

Bibliography

Carr, R.: Fundus flavimaculatus. Arch. Ophthalm., 74:163-168, 1965.

FRANÇOIS' CLOUDY CENTRAL DYSTROPHY

Synonym: *Dystrophie Nuegeuse Centrale.*

Ocular Findings: Bilateral central corneal dystrophy.

Central third of corneas are occupied by small grayish snowflake opacities which cover approximately the whole pupillary area. They show no definite structure or limits. They are denser near Descemet's membrane and fewer anteriorly and toward the periphery.

The peripheral area is always perfectly intact.

The sensitivity of the cornea is normal.

Vision is normal and there are no subjective symptoms.

This dystrophy has no relation to any other ocular defect.

May appear at any age and progresses extremely slow.

Etiology: Hereditary.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FRANÇOIS' SPECKLED DYSTROPHY

Synonym: *Dystrophie Mouchetée.*

Ocular Findings: Small dot-like opacities throughout the corneal stroma, visible only by slit-lamp.

Opacities are bilateral and identical on both sides.
Epithelium, Bowman's, Descemet's, and the endothelium are intact.
Sensitivity of the cornea is normal.

Associated Clinical Findings: Not associated with any general diseases
or any other ocular defect.
Appears to be congenital.
Does not progress with age.

Etiology: Familial. Regular autosomal dominant.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FRANKL-HOCHWART SYNDROME

Synonym: *Pineal-neurologic-ophthalmic Syndrome*.

Ocular Findings: Limitation of upward gaze.
Papilledema.
Concentric field constriction.

Associated Clinical Findings: Bilateral deafness.
Ataxia.
Hypopituitarism.

Bibliography

Tassman: *The Eye Manifestations of Internal Diseases*, 3rd ed. C. V. Mosby, St. Louis, Mo., 1951.

FREEMAN-SHELDON SYNDROME

Synonym: *Cranio-carpo-tarsal Dystrophy*.

Ocular Findings: Enophthalmos.
Epicanthus.
Hypertelorism.
Strabismus.

Associated Clinical Findings: Flat face with long philtrum and small mouth.

Ulnar deviation of the hands with contraction.

Clubfoot.

Normal intelligence.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

FRENKEL'S SYNDROME

Synonym: *Ocular Contusion Syndrome.*

Ocular Findings: Traumatic mydriasis, iridoplegia, hyphema, and iridodialysis.

Subluxation of the lens.

May result in peripheral retinal degeneration resembling atypical retinitis pigmentosa.

Etiology: Blunt trauma to the anterior segment of the globe.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

FRIEDREICH'S SYNDROME

Synonym: *Friedreich's Hereditary Ataxia.*

Ocular Findings: Nystagmus.

Occasional external ophthalmoplegia.

Associated Clinical Findings: Cerebellar ataxia.

Dysmetria.

Loss of deep tendon reflexes.

Adiadochokinesis.

Dysarthria.

Appears in children between eight and fifteen years of age.

Etiology: Familial. Simple recessive inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FROBOESE'S SYNDROME

Synonym: *Myelin Neuromatosis*.

Similar Condition: *von Recklinghausen's Disease*.

Ocular Findings: Non-painful neurocytoma of the ocular mucous membranes.

Hypertrophic corneal nevi have been reported.

Associated Clinical Findings: Non-painful neurinoma or neurocytomas of the mucous membranes of the lips, tongue, and esophagus.

Facial lentiginosis.

Skull deformations.

Seen in infancy.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

FRÖHLICH'S SYNDROME

Synonyms: *Adiposogenitalism, Adiposogenital Dystrophy*.

Ocular Findings: Bitemporal hemianopsia or scotomas.

Impairment of dark adaptation.

May have papilledema.

Associated Clinical Findings: Adiposity of juvenile or female type.

Genital hypoplasia.

Occasionally mental infantilism.

Pathologic Etiology: This is a hypothalamico-pituitary disorder interfering with centers regulating hunger and disturbing the gonadotropic function of the pituitary.

Craniopharyngeal tumors, suprasellar lesions, and occasionally, hypophyseal lesions such as chromophobe adenoma have been identified.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

FUCHS' DELLEN

Synonym: *Fuchs' dimples*.

Ocular Findings: Small depressions in the corneal stroma usually occurring near the periphery. They occur in corneal degenerations.

Fuchs' dellen or dimples are to be differentiated from Fuchs' crypts which are excavations in the pupillary zone of the iris which are lacking in surface endothelium so that aqueous fluid can get quickly in and out of the iris.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

FUCHS' DISEASE

Synonym: *Fuchs' Gyrate Atrophy of the Choroid and Retina*.

Ocular Findings: Slowly progressive gyrate atrophy of the choroid, the pigment epithelium, and the retina.

The fundus appears white but the macula and the optic papilla appear normal.

Hemeralopia.

Diminished vision.

Absent electroretinographic response.

Pathology: Adler's anomaly (azurophilic granulations) appearing in the polymorphonuclear leucocytes, basophiles and eosinophiles.

Etiology: Familial. Rare.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FUCHS' DYSTROPHY

Synonym: *Fuchs' Endothelial-epithelial Dystrophy*.

Ocular Findings: Hyaline, wart-like deposits on Descemet's membrane producing endothelial defects with edema and thickening of the epithelium, edema of the corneal stroma and degeneration of Bowman's membrane.

May be accompanied by pigmentation on the endothelium.

Corneal sensitivity is diminished.

May be unilateral or bilateral.

Usually begins in the fourth or fifth decade.

Affects women more than men by a ratio of three to one.

Etiology: Familial. Due to an irregularly dominant gene with incomplete penetrance.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

FUCHS' SIGN

Ocular Findings: Decreased mobility of the upper lid.

If the examiner grasps the upper lid and tries to pull it down, he feels a resistance more marked than that in normal subjects.

This sign is felt to be due to an increased tonus of the superior tarsal muscles.

Other eye signs of Graves' disease are frequently present.

Associated Clinical Findings: Systemic signs of hyperthyroidism may be present.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

FUCHS' SUPERFICIAL MARGINAL KERATITIS

Ocular Findings: Appears as a band of superficial infiltrates around the corneal periphery.

Central area of the cornea remains clear.

May be limited to one or two segments, or may be circumferential.

Develops slowly into shallow ulcerations with irregular scalloped edges.

Progresses to form a ring ulcer, then becomes vascularized as it progresses into the cornea producing pseudopterygia.

Typically bilateral.

Vision is not seriously impaired until late.

Usually occurs in elderly patients.

May be extremely painful.

Etiology: Obscure.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

FUCHS' SYNDROME

Synonyms: *Fuchs' Heterochromic Cyclitis*, *Fuchs' Complicated Heterochromia*.

Similar Condition: *See also Posner-Schlossman Syndrome.*

Ocular Findings: Mild recurrent anterior uveitis, usually unilateral.

Depigmentation and atrophy of the affected iris.

Tiny irregular keratic precipitates spreading over the entire posterior surface of the cornea.

Deterioration of the vitreous body with numerous floaters.

Posterior subcapsular saucer-shaped cataract (secondary cataract).

Ciliary injection is usually absent.

May be complicated by mild chronic glaucoma.

Appearance of a filiform pathognomonic hemorrhage in the angle when the anterior chamber is punctured (Aurruga's Sign).

Associated Clinical Findings: May be associated with facial hemiatrophy on the ipsilateral side.

Pathologic Etiology: Felt by some to be a heredodegenerative disease with disturbance of the sympathetic nervous system.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Goldberg, M. F.; Erozan, Y. S.; Duke, J. R.; and Frost, J. K.: Cytopathologic and histopathologic aspects of Fuchs' heterochromic iridocyclitis. *Arch. Ophthalmol.*, 74:604-609, 1965.

Kimura, S. J., Hogan, M. J., *et al.*: Fuchs' syndrome of heterochromic cyclitis. *A.M.A. Arch. Ophthalmol.*, 54:179-186, 1955.

FUCHS' SYNDROME

Synonym: *Mucocutaneous Ocular Syndrome*.

Ocular Findings: Severe conjunctivitis.
May have anterior uveitis.

Associated Clinical Findings: Headache.
Fever.
Cyanosis.
Ulcers of the mucous membranes.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

Thies, O.: The mucocutaneous ocular syndrome of Fuchs'. *Amer. J. Ophthalmol.*, 35:749, 1952.

FULLER-ALBRIGHT SYNDROME

Synonyms: *Osteitis Fibrosa Cystica Disseminata, Albright-Fuller Syndrome, McCune-Albright Syndrome*.

Ocular Findings: Proptosis.
Papilledema.

Optic atrophy.

Visual field changes secondary to optic atrophy.

Associated Clinical Findings: Osteitis fibrosa cystica.

The cystic bony changes are usually unilateral and involve the pelvic bones and lower extremities most frequently.

Present clinically with spontaneous fractures.

Brownish skin pigmentations appear on the same side as the bone lesions and vary from freckles to large patches.

Endocrine dysfunction which presents clinically as early menarche, adolescent external genitalia and breast enlargement (precocious puberty).

Seen more frequently in female children.

Bibliography

Albright, F.; Butler, A. M.; Hampton, A. D.; *et al.*: Syndrome characterized by osteitis fibrosa disseminata, areas of pigmentation, and endocrine dysfunction, with precocious puberty in females; report of five cases. *New Eng. J. Med.*, 216:727-746, 1937.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

G

GÄNSSLEN'S SYNDROME

Synonym: *Familial Hemolytic Icterus.*

Ocular Findings: Increased interpupillary distance.

Epicanthus.

Iris colobomas.

Associated Clinical Findings: Splenomegaly.

Polydactyly.

Deformities of the ears.

Hypogenitalism.

Pathology: X-rays show osteoporosis.

Etiology: Familial recessive. Occurs mainly in Caucasians.

Bibliography

Gänsslen, M.: Hemolytic jaundice. *Arch. Klin. Med.*, 140:210-226, 1926.

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

GARCIN'S SYNDROME

Ocular Findings: Unilateral ophthalmoplegia externa.
Visual field defects may be found.

Associated Clinical Findings: Defects of smell, unilateral.
Unilateral trigeminal paresis.
May have pareses of the ninth, tenth, eleventh, and twelfth nerves.
May have deafness, unilateral.

Pathologic Etiology: Nasopharyngeal, or brain stem neoplasm, or meningitis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

GASPERINI'S SYNDROME

Ocular Findings: Nystagmus.
Conjugate gaze pareses.
Sixth nerve paresis, unilateral.

Associated Clinical Findings: Unilateral paresis of the fifth and seventh nerves.
Deafness.
Contralateral hemiplegia.

Pathologic Etiology: Pontine lesions.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

GAUCHER'S DISEASE

Ocular Findings: Thickened conjunctiva resembling pinguecula in the palpebral fissure with a brownish discoloration.
Associated fatty degeneration of the cornea.
May have retinal hemorrhages.
May have associated paralytic strabismus.

Associated Clinical Findings: Dysphagia.

Progressive spasticity.

Brownish discoloration of the skin.

Spontaneous fractures.

In children may be acute and rapidly fatal.

In adults it is usually chronic.

Pathology: Accumulation of kerafin in the RE cells of the liver, spleen, and bones with hepatosplenomegaly.

Etiology: Familial. Irregular dominant.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

GAULE'S SPOTS

Ocular Findings: Sharply circumscribed areas of degeneration in the superficial layers of the cornea.

Resembles recurrent erosions of the cornea, with slight decrease in corneal sensitivity.

Characteristically seen in neuroparalytic keratitis.

Are more central than Fuchs' dellen.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

GERLIER'S DISEASE

Synonyms: *Kubisagari, Epidemic Paralyzing Vertigo.*

Ocular Findings: Ptosis.

Diplopia.

Transient ocular muscle pareses.

Associated Clinical Findings: Paroxysmal attacks of vertigo and muscle weakness.

Occurs in summertime and may be related to certain types of farm work around animals.

Gerlier's disease is limited to Switzerland and northern Japan. The attacks, which are often precipitated by fatigue, usually last about ten minutes, and may recur daily, or more frequently. Complete recovery occurs when cold weather sets in.

Etiology: Unknown.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

GERSTMAN'S SYNDROME

Synonym: *Angular Gyrus Syndrome*.

Findings: The patient is unable to name the fingers of either hand or tell the difference between the right and left hands.

Patients with Gerstman's angular gyrus syndrome may also have agraphia, acalculia, or both.

"Congenital reading disability" may actually be Gerstman's.

Pathologic Etiology: Usually caused by a thrombosis of the middle cerebral artery with a "left sided" lesion in the angular gyrus.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

GIFFORD'S SIGN

Ocular Findings: Difficulty everting the upper lid due to tonic spasm of the smooth muscles of the lid.

Occurs in about one-half of patients with thyrotoxicosis.

May be associated with thickening of the upper lid.

Associated Clinical Findings: Graves' Disease.

Increased basal metabolism and other systemic signs of thyrotoxicosis.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

GILLES DE LA TOURETTE SYNDROME

Ocular Findings: Blepharospasm.

Associated Clinical Findings: Facial spasms.

Attacks of aggressive anger, characterized by the use of abusive language, and dirty speech.

Pathology: This is felt to be a functional disease.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

GLOBE'S SIGN

Ocular Findings: Lid lag on upward gaze.

Frequently have other ocular signs of hyperthyroidism.

Associated Clinical Findings: Graves' Disease.

May have tachycardia, trembling, and increased basal metabolism.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

GODTFREDSSEN'S SYNDROME

Synonym: *Cavernous Sinus-neuralgia Syndrome*.

Ocular Findings: Unilateral ophthalmoplegia, usually involving the sixth cranial nerve.

Associated Clinical Findings: Anesthesia or neuralgia in the area of the trigeminal nerve.

Paralysis of the hypoglossal nerve on the ipsilateral side.

Pathologic Etiology: Usually caused by invasion of the cavernous sinus by tumor.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
- Godtfredsen, E.: Ophthalmo-neurological symptoms in malignant nasopharyngeal tumours. *Brit. J. Ophthal.*, 31:78-100, 1947.

GOLDENHAR'S SYNDROME

Synonym: *Oculoauriculovertebral Syndrome*.

Ocular Findings: Epibulbar dermoid and/or lipodermoids (usually bilateral).

Upper lid colobomas may be present.

Other congenital bulbar anomalies have been reported.

Associated Clinical Findings: Accessory auricular appendages and pretragal blind fistulas.

Facial microsmia.

Mild malar hypoplasia with recession of the chin and nostril atresia may be present.

Mild malar hypoplasia with recession of the chin and nostril atresia
Vertebral anomalies (usually diagnosed by x-ray).

Etiology: Congenital. Hereditary.

Bibliography

- Goldenhar, M.: Associations malformatives de l'oeil et de l'oreille, en particulier le syndrome dermoid épibulbaire-appendices auriculaires-fistulae auris congenita et ses relations avec la dysostose mandibulo-faciale. *J. de Génét. hum.*, 1:243, 1952.
- Sugar, H. S.: The oculoauriculovertebral dysplasia syndrome of Goldenhar. *Amer. J. Ophthal.*, 62:678, 1966.

GOPALAN'S SYNDROME

Synonym: "*Burning Feet Syndrome*."

Ocular Findings: Nutritional amblyopia.

Central scotomas.

Associated Clinical Findings: Aching foot paresthesias.

Sweating of the feet.

Etiology: B avitaminosis. Recorded primarily among prisoners of war.
May also be seen in diabetes.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

GOPPERT'S GALACTOSEMIC SYNDROME

Ocular Findings: Bilateral cataracts in infants.

Associated Clinical Findings: Galactosemia.
An assimilation defect of infancy.

Pathologic Etiology: A congenital metabolic disorder.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

GORLIN-CHAUDHRY-MOSS SYNDROME

Ocular Findings: Microphthalmia.
Oblique palpebral fissures.
Inability to close the lids fully.
Resultant exposure keratitis and corneal ulcerations followed by scarring.
Limited upward gaze.

Associated Clinical Findings: Craniofacial dysostosis.
Hypertrichosis.
Patent ductus arteriosus.
Dental anomalies.

Etiology: Hereditary.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.
Gorlin, R. J.; Chaudhry, A. P.; and Moss, M. L.: Craniofacial dysostosis, patent

ductus arteriosus, hypertrichosis, hypoplasia of labia majora, dental and eye anomalies—a new syndrome? *J. Pediat.*, 56:778-785, 1960.

GOUGEROT-SJÖGREN SYNDROME

Synonym: *Sjögren's Syndrome.*

Ocular Findings: Blepharoconjunctivitis and keratoconjunctivitis.

Conjunctiva is dry and soft to the touch.

Viscous secretions may be found in the cul-de-sacs.

Filamentous keratitis.

Recurrent corneal ulcerations.

Diminished lacrimal secretions.

Associated Clinical Findings: Xerostomia.

Buccal ulcerations.

Atrophic rhinitis.

Diarrhea.

Dry and ichthyotic skin due to absence of sudoral secretion.

Occurs mainly after third decade.

Affects females more than males.

Associated with rheumatoid arthritis.

Etiology: Familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

GRADENIGO'S SYNDROME

Synonym: *Lannois-Gradenigo Syndrome.*

Ocular Findings: Diplopia due to abducens palsy.

Pain around the eye.

Photophobia.

Lacrimation.

Reduced corneal sensitivity.

Associated Clinical Findings: Ipsilateral facial pain.

Otitis media.

Mastoiditis.

May have deafness on the ipsilateral side.

Pathologic Etiology: Usually produced by meningitis at the tip of the petrous bone.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

GRAEFE'S SIGN

Ocular Findings: Lid retraction on downward gaze.
Exophthalmos.

Associated Clinical Findings: Noncongestive dysthyroid disease.

This is a relatively insignificant, but interesting, feature in the differentiation of the congestive from the non-congestive type of dysthyroid exophthalmos.

In the non-congestive type the lid retraction is greatest on downward gaze, the typical Graefe sign. In the congestive type the retraction, when present, is greatest on upward gaze.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

GRAVES' DISEASE

Synonyms: *Basedow's Disease, Parry's Disease, Thyrotoxicosis, Hyperthyroidism.*

Ocular Findings: Unilateral or bilateral exophthalmos.

Any of the following eye signs, alone or in combination: Ballet's, Boston's, Cowen's, Dalrymple's, Enroth's, Fuchs', Gifford's, Globe's, Graefe's, pseudo-Graefe's, Jellineck's, Joffroy's, Knies', Lowe's, Moebius', Parry's, Rosenbach's, Sainton's, Saker's, Stellwag's, von Graefe's, Wilder's.

Associated Clinical Findings: Tachycardia.
Trembling.

Occasionally emaciation in severe cases.
Increased basal metabolism.
Frequently pretibial edema.

Pathologic Etiology: Condition induced by hyperplasia of the thyroid parenchyma and excessive secretion of its hormonal substance. Many of its eye signs are felt to be due to tonic contraction or hyperplasia of the smooth muscles of the lids and orbit.

Etiology: Unknown, but a strong familial tendency to the disease has been observed.

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- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

GREELEY'S CONJUNCTIVITIS

Synonyms: *Pharyngoconjunctival Fever, Beal's Conjunctivitis.*

Ocular Findings: Acute follicular conjunctivitis.
Transient keratitis.
Scanty mononuclear exudate in conjunctival sac.

Associated Clinical Findings: Pharyngitis.
Preauricular lymphadenopathy.
Fever.
Incubation period five to seven days.
Subsides in two to three weeks.

Etiology: Infection traced to swimming pools and droplet transmission. Type three adenovirus incriminated.
Received this eponym following a dramatic epidemic through the medium of a public swimming pool in Greeley, Colorado.

Bibliography

- Sorsby, A.: *Modern Ophthalmology*. Butterworth, Inc., Washington, D.C., 1963.
Theodore, F.: Pharyngoconjunctival fever and epidemic keratoconjunctivitis. *Eye Ear Nose Throat Monthly*, 45:94-95, 1966.

GREGG'S SYNDROME

Synonym: *Maternal Rubella Syndrome.*

Ocular Findings: Congenital nuclear cataracts.

Associated Clinical Findings: Congenital heart disease (commonly patent ductus).

Deafness.

Microcephaly.

Mental retardation.

Pathologic Etiology: Found in babies born of mothers who had contracted rubella in the early months of pregnancy.

Bibliography

Cooper, L.: German measles. *Sci. Amer.*, 215:30-37, 1966.

Dudgeon, J. A.; Butler, N. R.; and Plotkin, S. A.: Further serological studies on the rubella syndrome. *Brit. Med. J.*, 2:155-160, 1960.

Edit: Pathogenesis of rubella cataract. *Arch. Ophthalm.*, 73:761-763, 1965.

Gregg, N.: *Trans. Ophthalm. Soc. (Aust.)*, 3:35, 1941.

Zimmerman, L. E. and Fort, R. L.: Congenital malformations of the eye; some recent advances in knowledge of the pathogenesis and histopathological characteristics. *JAMA*, 196:684-692, 1966.

GREIG'S SYNDROME

Synonym: *Ocular Hypertelorism Syndrome.*

Ocular Findings: Wide spacing of the orbits.

Enophthalmos.

Epicanthus.

Deformities of the lids and brows.

Bilateral sixth nerve paralysis.

High astigmatism.

Secondary optic atrophy.

Convergent strabismus.

Associated Clinical Findings: Skull malformations.

Mental impairment.

Etiology: Probably familial.

Bibliography

- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Meisenbach, A. E., Jr.: Bilateral paralysis of external rectus muscle in hypertelorism; report of case with convergent strabismus. *Amer. J. Ophthalm.*, 33:83-87, 1950.

GRIGNOLO'S SYNDROME

Ocular Findings: Iritis.
Hypopyon.

Associated Clinical Findings: Erythema exudativum multiforme.
Spondyloarthritis.
Ankylosis.
Characterized by remissions and exacerbations.

Bibliography

- von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

GROENBLAD-STRANDBERG SYNDROME

Synonyms: *Angeoid Streaks of the Retina, Systemic Elastodystrophy.*

Ocular Findings: Ruptures in Bruch's membrane, with resultant grayish irregular streaks in the retina.
May have macular hemorrhages and transudates.

Associated Clinical Findings: Peripheral vascular disturbances.
Gastrointestinal hemorrhages.
Myocarditis and aortitis.
Premature arteriosclerosis.
Secondary degeneration of the elastic tissues throughout the body.
May be associated with pseudoxanthoma elasticum (see Ehlers-Danlos syndrome).

Etiology: Familial.

Bibliography

- Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Groenblad, E.: Angoid streaks—pseudoxanthoma elasticum; vorläufige mitteilung. ACTA. Ophthal., 7:329, 1929.

GROENOUW'S DYSTROPHY TYPE I

Synonyms: *Groenouw's Granular Corneal Dystrophy, Fleischer's Dystrophy, Familial Granular Dystrophy.*

Ocular Findings: Small, whitish, irregular, often flattened dot-like opacities in the superficial layers of the cornea.

Develop in the central area of the cornea.

The opacities appear to form a granular disc localized in the anterior layers of the cornea.

The deeper layers remain intact and the periphery remains transparent.

Corneal sensitivity is reduced.

Pathology: Degeneration of the anterior stromal cells of the cornea.

Etiology: Familial. Dominant inheritance. Appears in early childhood.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Sornson, E. T.: Granular dystrophy of the cornea. An electron microscopic study. Amer. J. Ophthal., 59:1001-1007, 1965.

GROENOUW'S DYSTROPHY TYPE II

Synonyms: *Spotted Dystrophy, Fehr's Dystrophy.*

Ocular Findings: Central cornea shows diffuse flecking and irregularity. Diminished vision.

The whitish opacities, at first small and indistinct and of varying thickness, appear first superficially and lift up the epithelium of the cornea.

Usually begins about age five to nine.

By the second decade small gray spots also appear near Descemet's both at the periphery and centrally.

In the final stages, the opacities are grouped in irregular masses of various sizes and shapes in all layers of the cornea.

Corneal sensitivity is reduced.
May have recurrent erosions.
Patient may be blind by the fourth decade.

Etiology: Familial. Autosomal recessive type inheritance.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. *Arch. Ophthalm.*, 74:516-520, 1965.

GRUBER'S SYNDROME

Synonym: *Splanchnocystic Dysencephaly*.

Ocular Findings: Hypertelorism.

Shallowness of the orbits.

Exophthalmos.

Associated Clinical Findings: Dysostosis of the skull.

Polydactyly.

Status dysraphicus.

Genital dysplasia.

Multiple cystic formations in such internal organs as the liver, pancreas, kidney, and ovary.

Etiology: Familial.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

GRUNER-BERTOLOTI SYNDROME

Synonym: *Combined Parinaud's and Von Monakow's Syndrome*.

Ocular Findings: Supranuclear paralysis of conjugate upward movements of the eyes without paralysis of convergence.

Associated Clinical Findings: Syringomyelic sensory changes.

Fibrillar tremors.

Associated hemiplegic paralysis.

Pathology: The lesion may involve the midbrain; more particularly the superior part of the cerebral peduncles.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

GUIAT'S SIGN

Ocular Findings: Tortuosity of the retinal veins.

Pathologic Etiology: Guiat's sign is considered an early sign of retinal arteriolar sclerosis.

GUILLAIN-BARRÉ SYNDROME

Synonyms: *Landry's Paralysis, Acute Polyneuritis.*

Ocular Findings: Paralytic ectropion of the lower eyelids due to facial nerve paralysis.

May have complete external ophthalmoplegia.

Pupils may be mydriatic.

May have papilledema or optic atrophy.

Associated Clinical Findings: Polyneuritis.

Facial diplegia.

Absent tendon reflexes.

Pareses of the extremities.

Pathology: Degeneration of the anterior horn cells.

Etiology: Unknown. Usually occurs from age fifteen to fifty. About 20% mortality

Bibliography

Ford, F. R. and Walsh, F. B.: Guillain-Barré syndrome (acute infective polyneuritis) with increased intracranial pressure and papilledema. *Hopkins Hosp.*, 73:391-395, 1943.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957

GUNN'S PUPILLARY SIGN

Synonyms: *Marcus Gunn Pupil Sign, Greeves' Sign, Afferent Pupil Sign.*

Ocular Findings: On illumination of the affected eye, the pupil dilates after a short, slight contraction, an apparently paradoxical reaction.

Pathologic Etiology: This is a sign of retrobulbar neuritis.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

GUNN'S SIGN

Synonym: *A-V Nicking, see also Salus Sign.*

Ocular Findings: Compression (nicking) of the veins at the points of crossing of the retinal arterioles.
Vein may be concealed by the artery.

Pathologic Etiology: Hypertension.

GUNN'S SYNDROME

Synonyms: *Marcus Gunn's Syndrome, Jaw-Winking Syndrome, Maxillopalpebral Synkinesia.*

Ocular Findings: Unilateral congenital ptosis with paradoxical elevation of the lid when the mouth is opened or mandible is moved to one side or the other.

Occasional palsy of external ocular muscles may be seen.

Pathology: Felt to be due to abnormal connections between the central innervation of the external pterygoid muscle and the levator superioris.

Etiology: May be autosomal dominant.

Bibliography

- Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.
- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
- Gunn, M.: Congenital ptosis with peculiar associated movements of the affected lid. *Trans. Ophthal. Soc. UK.*, 3:283-287, 1883.

H

HAAB'S SENILE MACULAR DEGENERATION

Ocular Findings: Punctate macular chorioretinal degeneration.

Pathology: Obliteration of the choriocapillaris.

HAAB-DIMMER DYSTROPHY

Synonyms: *Biber-Haab-Dimmer Corneal Dystrophy, Lattice Dystrophy.*

Ocular Findings: Lattice-like dystrophy of the corneal stroma.

Pathology: The stroma presents hyaline degeneration and absence of acid mucopolysaccharides as in granular dystrophy but as contrasted with macular dystrophy.

Etiology: Familial. Recessive.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
- Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. *Arch. Ophthal.*, 74:516-520, 1965.

HAENEL'S SYNDROME

Synonyms: *Haenel's Sign, Tabetic Ocular Anesthetic Syndrome.*

Ocular Findings: Absence of pain on firm pressure on or over the eyeballs in patients with tabes dorsalis.

This phenomenon is used as a diagnostic criterion in the late or chronic stage of neurosyphilis as it is known that an *increase* in pain sensation is commonly felt by pressure on the globe of the eye during the early period of tabes dorsalis.

Etiology: Syphilis.

Bibliography

- Duke-Elder, Sir S.: *Textbook of Ophthalmology*, Vol. 4. C. V. Mosby, St. Louis, Mo., 1949.
Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

HALLERMAN-STREIFF SYNDROME

Synonym: *Dyscephalia-mandibulo-oculo-facialis*.

Ocular Findings: Eyes are small.

Orbits may be small.

Strabismus.

May have cataracts.

Associated Clinical Findings: Small beaked nose, "bird face."

Sharp naso-frontal angle.

Ears are low-set.

Mouth is small and lips are thin.

Hypoplastic mandible.

Brachycephalic skull.

Diminished hair on scalp and patchy alopecia.

Etiology: Congenital. Familial.

Bibliography

- Donaldson, D.: *Stereo Atlas of External Diseases of the Eye*. C. V. Mosby, St. Louis, Mo., 1966.
Hoefnagel, D. and Benirschke, K.: *Dyscephalia-mandibulo-oculo-facialis*. *Arch. Dis. Child.*, 40:57-61, 1965.
Keitel, H., *et al.*: Peculiar versus diagnostic facies in pediatrics. *Lippincott's Med. Science*, 17:47-58, 1966.

HALLGREN'S SYNDROME

Ocular Findings: Pigmentary retinal degeneration.

Cataracts.

Occasional nystagmus.

Associated Clinical Findings: Vestibulo-cerebellar ataxia.

Deafness.

Mental deficiency.

Psychosis.

Etiology: Hereditary. Autosomal recessive.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

HAND-SCHÜLLER-CHRISTIAN SYNDROME

Synonym: *Xanthomatous Granuloma Syndrome.*

Ocular Findings: Exophthalmos, bilateral in about seventy per cent of cases.

Xanthelasma of the lids.

May have internal ophthalmoplegia.

May have nystagmus.

May have corneal degeneration in later stages.

May have papilledema followed by optic atrophy in severe cases.

Associated Clinical Findings: Diabetes insipidus.

Hepatosplenomegaly.

Osseous lesions of the membranous bones of the skull with growth retardation.

Pathology: Granulomatous involvement of the reticuloendothelial system.

Etiology: Possibly male sex-linked but is rarely familial.

Bibliography

Davison, C.: Xanthomatosis and central nervous system (Schüller-Christian syndrome). *Arch. Neurol. Psychiat.*, 35:629-630, 1936.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

HANHART'S SYNDROME

Ocular Findings: Superficial dendritic ulcerative lesions in the corneal epithelium. Normal corneal sensitivity is retained.

Associated Clinical Findings: Dyskeratosis palmo-plantaris.
Anomalies of the skin.

Etiology: Uncertain.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

HANSEN'S DISEASE

Synonym: *Leprosy*.

Ocular Findings: Severe and overwhelming tuberculoid anterior uveitis.
While it should be noted that a true leprous conjunctivitis probably does not exist as such, a keratoconjunctivitis from exposure is not at all uncommon.

Associated Clinical Findings: Leonine facies.
Systemic signs of leprosy.

Pathologic Etiology: Infection with mycobacterium leprae.

Bibliography

Datelines in Ophthalmology, Vol. 2., No. 1., 1965.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

HARADA'S SYNDROME

Synonym or Similar Syndrome: *Vogt-Koyanagi Syndrome*.

Ocular Findings: Bilateral uveitis.
Exudative choroiditis.
Serous retinal detachment.
Detachment may spontaneously reattach after several weeks; depigmentation and patches of chorioretinal scarring appear later.

Associated Clinical Findings: Alopecia.
Poliosis.
Vitiligo.
Hearing defects.

Headaches and signs of meningeal irritation.
Usually affects young adults.

Etiology: Unknown but may be viral.

Bibliography

- Cowper, A. R.: Harada's disease and Vogt-Koyanagi syndrome: Uneoencephalitis. A.M.A. Arch. Ophthal., 45:367-376, 1951.
Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

HARRIS' SYNDROME

Synonym: *Periodic Migraine.*

Similar Condition: *Horton's Syndrome.*

Ocular Findings: Tearing and excessive lacrimation.
Ciliary neuralgia.
Peri and supra-orbital headaches.

Associated Clinical Findings: Severe recurrent unilateral headaches beginning behind or above the eye and radiating to the neck and shoulder.
Usually begin in the morning.
Characterized by periodic recurrences.
Usually affect males under age forty.

Bibliography

- von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

HASSALL-HENLE BODIES

Synonym: *Hassall-Henle Warts.*

Ocular Findings: Hyaline excrescences pushing out between the cells of the endothelium, producing nodular thickenings at the periphery of Descemet's.

Seen as microscopic nodules or wart-like bumps of the endothelium.
They are comparable to drusen of the lamina vitrea of the choroid.

Hassall-Henle bodies are rarely seen before the age of twenty and increase in number with advancing age.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

HEBRA'S DISEASE

Synonyms and Similar Conditions: *Erythema Multiforme Exudativum*, *Stevens-Johnson Disease*.

Ocular Findings: Conjunctivitis which may be catarrhal (most common), purulent, or pseudomembranous (in severe cases).

Corneal ulcers may appear in the course of the disease.

Iritis.

Symblepharon in later stages.

Associated Clinical Findings: Cutaneous erythematous patches sometimes involving the face and mucous membranes.

Occasional severe stomatitis.

Historical Note: Stevens and Johnson, in 1922, described two severe cases of this disease, calling it, "eruptive fever with stomatitis and ophthalmia," classifying it as a "new" disease, hence the eponym for the more dramatic cases. Thus, if stomatitis is prominent it is labeled Stevens-Johnson disease.

Etiology: Possibly toxic or allergic.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

HEERFORDT'S DISEASE

Synonym: *Uveoparotid Fever*.

Ocular Findings: Bilateral granulomatous uveitis.

Associated Clinical Findings: Boeck's sarcoidosis.

Parotid gland inflammation.

Lymphadenopathy.

Low grade fever.
Occasional facial paralysis.
Females affected more than males.
Most frequent in young adults.

Etiology: Unknown. May be a manifestation of systemic sarcoidosis.

Bibliography

- Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.
Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Heerfordt, C. F.: Ueber eine Febris uneo-parotidea subchronica an der glandula parotis und der unea des auges lokalisiert und häufig mitparesen cerebros spinaler nerven kompliziert. Arch. Ophthal., 70:254-273, 1909.

HEIDENHAIN'S SYNDROME

Synonym: *Presenile dementia-cortical Blindness Syndrome*.

Ocular Findings: Rapid loss of vision with normal fundi (cortical blindness).

May have hemianopsia.

Associated Clinical Findings: Dysarthria.

Ataxia.

Athetoid movements.

Generalized rigidity.

Pathology: Autopsy findings show cortical degeneration especially marked in the occipital cortex.

Bibliography

- Meyer, A.; Leigh, D.; and Bagg, C. E.: A rare presenile dementia associated with cortical blindness (Heidenhain's syndrome). J. Neurol. Neurosurg. Psychiat., 17:129-133, 1954.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed., Williams and Wilkins, Baltimore, Md., 1957.

HELMHOLZ-HARRINGTON SYNDROME

Synonym: *Corneal Opacity-cranioskeletal Dysostosis Syndrome*.

Ocular Findings: Congenital dystrophic corneal opacities consisting of diffuse cloudiness.

Associated Clinical Findings: Mental retardation.

Limitation of movement of arms and legs.

Short, clawlike hands and feet.

Lumbar kyphosis.

Scaphocephalic head.

Seen in infancy and early childhood.

Etiology: Familial.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Helmholz, H. F. and Harrington, E. R.: Syndrome characterized by congenital clouding of cornea and by other anomalies. *Amer. J. Dis. Child.*, 41:793-800, 1931.

HENNEBERT'S SYNDROME

Synonym: *Luetic-otitic-nystagmus Syndrome*.

Ocular Findings: Attacks of spontaneous nystagmus and giddiness with exaggeration of the nystagmus when the column of air in the auditory meatus is compressed.

Associated Clinical Findings: Labyrinthine fistula. The increased nystagmus on air compression is diagnostic of labyrinthine fistula.
Vertigo.

Bibliography

Asherson, N.: Spontaneous nystagmus in congenital syphilis. *Arch. Dis. Child.*, 5:331-334, 1930.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

HENSY'S SIGN

Synonym: *Orbicularis Sign*.

Ocular Findings: This sign consists of the patient's inability to close his eye on the paretic side without closing his other eye.

Pathologic Etiology: Seen in facial palsy.

This is also seen occasionally in a definite "one eye dominance."

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

HERBERT'S PITS

Ocular Findings: Follicle remnants in the cornea in cases of trachoma. In healing trachoma, the corneal follicles rupture or cicatrize and the filling of the defect by optically clear epithelium leads to the production of apparently clear cavities. And so, Herbert's pits are actually depressions left as the trachoma follicles resolve.

Etiology: Trachoma.

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Duke-Elder, Sir S.: *Textbook of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1949.
Internat. Ophth. Clin. Vol. 2. No. 3. p. 596.

HERBERT'S ROSETTES

Ocular Findings: In trachomatous keratoconjunctivitis, an early change leading to the formation of pannus.

As the capillaries advance into the cornea, a fine lymphocytic infiltration appears around them. These may become aggregated into follicles in the limbal area or in the peripheral cornea, showing up as slightly elevated translucent nodules surrounded by a fine capillary network (Herbert's rosettes).

Etiology: Trachoma.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

HERING'S AFTER-IMAGE

Synonym: *Positive After-image*.

Ocular Findings: Positive after-image, having the same qualities of the original stimulus or image.

They are produced by the first stimulus and are therefore called "original" or "real" after-images. For instance, if one looks at a white light, the positive after-image will appear white.

Opposite to Purkinje's negative after-image or Bidwell's ghost.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

HERRICK'S SYNDROME

Synonyms: *Dresbach's Syndrome, Sickle Cell Disease.*

Ocular Findings: Conjunctival capillary stasis.

Secondary glaucoma.

Vitreous hemorrhages.

Complicated cataract.

Hemorrhagic and exudative retinopathy.

Retinitis proliferans.

Associated Clinical Findings: Sickle cell anemia.

Hepatosplenomegaly.

Moderate jaundice.

Seen almost exclusively in Negroes.

Pathology: Sickled erythrocytes. Hyperplastic bone marrow.

Etiology: Inherited. Dominant transmission in Negroes.

Bibliography

Goodman, G.; Sallmann, L. von; and Holland, M. G.: Ocular manifestations of sickle-cell disease. *A.M.A. Arch., Ophthal.*, 58:655-682, 1957.

Margolies, M. P.: Sickle cell anemia; composite study and survey. *Medicine*, 30:357-443, 1951.

HERTWIG-MAGENDIE SYNDROME

Synonyms: *Skew Deviation, Hertwig-Magendie Squint.*

Ocular Findings: In this rare phenomenon one eye deviates downward and inward, the other eye upward and outward. It may be considered as a vertical divergence of the horizontal midplanes of the two eyes.

The picture of Hertwig-Magendie squint may be imitated by a paresis of the elevators of one eye and the depressors of the other eye or by alternating hyperphoria. They can be differentiated by the relative incomitancy of the paretic type on testing versions.

Pathologic Etiology: Skew deviation has been seen with cerebellar tumors, epidemic encephalitis, and after prefrontal lobotomy.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

HILDING'S SYNDROME

Synonym: *Uveo-arthro-chondral Syndrome*.

Ocular Findings: Chronic uveitis.

Associated Clinical Findings: Dystrophy of the cartilage of the ears and the long bones, especially the ribs.
Joint subluxation.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

HIRSCHBERG'S SIGN

Synonym: *Ocular Rubeola Sign*.

Ocular Findings: Lesions similar to Koplic spots appearing on the caruncle and occasionally on the conjunctiva one or two days before they appear on the buccal mucosa.

May precede or be accompanied by photophobia.

Pathologic Etiology: Pathognomonic of measles (rubeola).

Bibliography

Allen, J. H.: Ocular lesions associated with other viral diseases. *Internat. Ophthalm. Clin.*, 4:311-324, 1964.

HIRSCHBERG'S SIGN

Synonym: *Hirschberg's Light Reflex.*

Ocular Findings: This is a method of estimation of the angle of squint deviation.

Every millimeter of decentration from the center of the pupil in the deviating or non-fixing eye is approximately equivalent to fifteen prism diopters.

Tested best by having the patient attempt binocular fixation on a bright point light source.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

HODGKINS' DISEASE

Synonym: *Lymphogranulomatosis.*

Ocular Findings: Uveitis.

Associated Clinical Findings: Systemic Hodgkins' Disease, with progressive enlargement of lymph nodes and spleen and liver.

In later stages, fever, anemia and cachexia are present. May have pruritis.

Pathology: Involvement of the eye in Hodgkins' disease is rare, but nodules have been removed from the orbit which have shown the typical histologic picture.

Hodgkins' disease has been reported associated with torulosis, and while the association is not clear, it is too frequent to be a mere coincidence.

Etiology: Unknown.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

HOLTERMÜLLER-WIEDEMANN SYNDROME

Ocular Findings: Optic atrophy.

Blindness.

Orbital anomalies.

Associated Clinical Findings: Congenital hydrocephalus.

"Cloverleaf skull."

Progressive mental deterioration.

Spina bifida.

Abnormal smallness of the limbs.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

HORNER'S SYNDROME

Synonym: *Bernard-Horner Syndrome.*

Ocular Findings: Usually unilateral pupillary constriction but with reaction to light and accommodation.

Lid reflex is retained.

The pupil fails to dilate with cocaine, but will dilate with adrenalin, which differentiates it from a spastic miosis.

The palpebral fissure is narrowed with the upper lid drooping (ptosis) and slight elevation of the lower lid, secondary to interruption of smooth muscle innervation.

Apparent enophthalmos.

Excessive tearing may be seen.

Associated Clinical Findings: Ipsilateral anhidrosis of the face and neck.

Occasional facial hemiatrophy.

Pathology: Interruption of the sympathetic chain in the cervical region.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Jaffe, N. S.: Horner's syndrome. *Amer. J. Ophthal.*, 34:1182-1183, 1951.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

HORNER-TRANTAS SPOTS

Synonym: *Trantas Spots.*

Ocular Findings: White, chalky concretions appearing as small white specks in the area of tarsal papillary hypertrophy.

Seen in the later stages of allergic or vernal keratoconjunctivitis.

Etiology: Ocular allergy.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

HORTON'S SYNDROME

Synonym: *Histamine Cephalalgia Syndrome.*

Ocular Findings: Hyperemia of the conjunctiva.
Lacrimation.

Associated Clinical Findings: Brief, recurrent, violent, unilateral headaches involving the temple, eye, orbit, and the side of the neck.
May also have rhinorrhea.
May have increased skin temperature over the affected area.
Occurs chiefly in males over 40 years of age.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Horton, B. T.: Use of histamine in treatment of specific types of headaches. *JAMA*, 116:377-383, 1941.

HORTON-MAGATH-BROWN SYNDROME

Ocular Findings: Ischemic optic neuritis.
Central retinal vein occlusion frequently reported.
Eye muscle pareses.

Associated Clinical Findings: Temporal arteritis with low skin temperature in the involved area.

Headaches (like Horton's histamine headaches).

Usually seen in older males.

Accompanied by progressive weakness.

Hypochromic anemia.

Pathology: Diagnosis is by arteriectomy with demonstrable periarteritis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

HOUSSEY'S PHENOMENON

Findings: Reversibility of diabetic retinopathy when pituitary function is lost. Clinically, this may occur in cases of pituitary necrosis from infarct, etc. Surgically it may follow hypophyseal section, and this is the rationale for the procedure.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 7. C. V. Mosby, St. Louis, Mo., 1962.

HUDSON-STAHLI LINE

Synonyms: *Hudson's Line, Stahli's Line.*

Ocular Findings: A wavy pigment line about one-half millimeter wide on the lower cornea, at about where the lids meet.

Appears as a linear horizontal line, brown or greenish in color, crossing the cornea at the junction of the middle and lower thirds. There is often a slight convexity of the line directed inferiorly.

Occurs usually in older people but may appear in any age group.

Pathology: Occurs in cases of epithelial degeneration. It has been shown to be a deposit of hemosiderin and may or may not be related to tears in Bowman's membrane.

Bibliography

Gass, J. D.: The iron lines of the superficial cornea. *Arch. Ophthal.*, 71:348-358, 1964.
Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

HUGHLINGS-JACKSON SIGN

Ocular Findings: Pupillary dilation with cocaine in cases of levator ptosis. This is the pharmacologic response in differentiation between levator ptosis and sympathetic ptosis.

In sympathetic ptosis the pupil is not dilated with cocaine, but reacts with normal dilation to homatropine.

In levator ptosis, the pupil dilates with cocaine.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

HUNTER'S SYNDROME

Synonym: *Mucopolysaccharidosis II*.

Ocular Findings: Corneal clouding seen as a deep stromal haze by slit lamp.

Associated Clinical Findings: Superficially Hunter's syndrome resembles Hurler's syndrome in that dwarfing, stiff joints, hepatosplenomegaly, and gargoyle-like facies are commonly observed. However, the clinical severity of all abnormal features is less than in Hurler's syndrome and the life span may extend through the fourth decade.

Absence of corneal clouding in childhood is one of the most reliable differences between this syndrome and Hurler's.

Corneal clouding does develop as the patient gets older and presents as a deep stromal haze by slit-lamp in adults.

Etiology: Inherited as an X-linked recessive trait.

Bibliography

Gills, J. A.; Hobson, R.; Hanley, W. B.; *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. *Arch. Ophthalm.*, 74:596-603, 1965.

Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. *Arch. Ophthalm.*, 74:516-520, 1965.

HUNTINGTON'S DISEASE

Synonym: *Huntington's Chorea.*

Ocular Findings: Vertical gaze palsy.
Strabismus.

Associated Clinical Findings: Personality changes.

Involuntary choreiform movements appearing first in the face, neck, and upper extremities, which are jerky, irregular, and stretching in character.

Usual onset in the fourth to the sixth decades.

Pathology: Degenerative changes in the cerebral cortex, in the caudate and lenticular nuclei.

Etiology: Hereditary.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

HURLER'S DISEASE

Synonyms: *Pfaundler-Hurler Syndrome, Gargoylism, Lipochondrodystrophy, Mucopolysaccharidosis I.*

Ocular Findings: Corneal clouding with lipid deposits in all layers except the epithelium and endothelium.

Corneal opacities may conceal concurrent retinal degeneration.

Occasionally optic atrophy may occur.

Unlike macular corneal dystrophy (Groenouw's II), irritative exacerbations are unlikely to occur.

Associated Clinical Findings: Dwarfism.

Gargoylism.

Hepatosplenomegaly.

Mental deficiency (initial normal development with subsequent regression into a state of mental deficiency).

Pathology: Disturbance of carbohydrate metabolism. The urine contains excessive mucopolysaccharides.

Etiology: Familial.

Bibliography

- Gills, J. A.; Hobson, R.; Hanley, W. B.; *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. Arch. Ophthalm., 74:596-603, 1965.
- Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. Arch. Ophthalm., 74:516-520, 1965.

HUTCHINSON'S DISEASE

Synonym: *Hutchinson and Tay's Choroidal Guttata.*

Ocular Findings: A form of senile cystic macular dystrophy.

It is marked by the presence near the posterior pole of light yellowish or tawny spots which look like little drops. Around these spots, there may be other larger ones, whiter and sometimes pigmented around the edges.

Pathology: Senile colloid degeneration. Probably through alterations of the lamina vitrea.

Bibliography

- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
- Krill, A. E. and Klein, B. A.: Flecked retina syndrome. Arch. Ophthalm., 74:496-508, 1965.

HUTCHINSON'S SYNDROME

Synonym: *Hutchinson's Triad.*

Ocular Findings: Interstitial keratitis.

The interstitial keratitis is a late manifestation of hereditary syphilis and produces vascularization and permanent opacities of the cornea. Interstitial keratitis may not appear until ten or more years after the disease is acquired.

Associated Clinical Findings: Dysacucia.
Teeth deformities.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

I

IWANOFF CYST

Synonym: *Blessig Cyst*.

Ocular Findings: Peripheral retinal degenerative cysts, occurring in the non-functional portion of the retina just behind the ora.

May extend five to seven millimeters back of the ora.

Usually appear after the age of thirty.

May predispose to retinal detachment, though it is usually considered a physiologic finding.

Pathologic Etiology: Degenerative changes in the outer layers of the retina.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

J

JACKSON'S SYNDROME

Similar Condition: *Avellis Syndrome*.

Ocular Findings: Miosis.

Ptosis.

Relative enophthalmos (Horner's Syndrome).

Associated Clinical Findings: Involvement of the hypoglossal nerve (paralysis of the tongue).

Pareses of the sternomastoid, palate and vocal cords.

Pathologic Etiology: Lesions of the medulla.

Does not show facial paralysis as you would find in pontine lesions.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

JACOB'S SYNDROME

Synonym: *Oculo-oro-genital Syndrome.*

Ocular Findings: Bulbar conjunctivitis.

Keratitis.

Optic atrophy.

Associated Clinical Findings: Stomatitis.

Glossitis.

Exfoliative dermatitis of the scrotum.

Aphthous ulcers of the buccal mucous membranes.

Pharyngitis.

Etiology: Vitamin A and B deficiency. Prognosis good with treatment.

Bibliography

Jacobs, E. C.: Oculo-oro-genital syndrome: Deficiency diseases. *Ann. Int. Med.*, 35:1049-1054, 1951.

JACOD'S SYNDROME

Synonyms: *Negri-Jacod's Syndrome, Petrosphenoidal Space Syndrome.*

Ocular Findings: Unilateral optic atrophy with amaurosis.

Ophthalmoplegia.

Ophthalmic branch trigeminal neuralgia.

Associated Clinical Findings: Thirty per cent of cases have cervical lymphadenopathy

Pathologic Etiology: Tumors of aneurysms in the petrosphenoidal space.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

Godtfredsen, E.: Ophthalmic-neurological symptoms in malignant nasopharyngeal tumours. *Brit. J. Ophthalm.*, 31:78-100, 1947.

JADASSOHN-LEWANDOWSKY SYNDROME

Similar Condition: *Zinsser-Thomson Disease.*

Ocular Findings: Corneal dystrophy.
Cataracts.

Associated Clinical Findings: Mental retardation.
Skin thick and horny.
Polykeratosis.
Alterations in the fingernails (onychogryposis).
More common in males.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

JAHNKE'S SYNDROME

Similar Condition: *See Sturge-Weber Syndrome.*

Ocular Findings: Choroidal angiomas.
May have heterochromia iridis without glaucoma.
Conjunctival telangiectasis.

Associated Clinical Findings: May have meningeal angiomas.

Etiology: Familial. May be simple recessive.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

JAYLE-OURGAUD SYNDROME

Synonym: *Ataxic nystagmus.*

Ocular Findings: Anterior or posterior internuclear ophthalmoplegia.
Paresis of conjugate lateral deviation.
Nystagmus.
The paralysis of the medial rectus in lateral gaze is associated with nystagmoid movements of the contralateral eye during the same lateral conjugate movement.

Bibliography

- Hugonnier, R. and Magnard, P.: Paralyse internucléaire antérieure. Bull. Soc. Ophthal. France, 58:265-268, 1958.
- Orlowski, W. J.; Slomski, P.; and Wojtowicz, S.: Bielschowsky-Lutz-Cogan syndrome. Amer. J. Ophthal., 59:416-430, 1965.

JEFFERSON'S SYNDROME

Synonym: *Cavernous Sinus (Forward Wall) Syndrome.*

Ocular Findings: Eye muscle pareses.

Associated Clinical Findings: In the frontal or forward wall type, the first division of the fifth nerve is affected.

If the causative lesion extends farther, the second division of the fifth may be involved.

If all divisions of the fifth are involved as well as the sixth (posterior wall type) it is called the Jefferson-Bonnet Syndrome.

Etiology: Aneurysms are the most frequent cause.

Bibliography

- von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

JELLINEK'S SIGN

Ocular Findings: Increased pigmentation of the outer surface of the closed eyelids.

Found infrequently in cases of Graves' disease.

Associated Clinical Findings: May have systemic signs of hyperthyroidism.

Increased basal metabolism.

Tachycardia.

Tremors.

Etiology: Unknown. Relationship to Graves' disease is unknown.

Bibliography

- Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

JOFFROY'S SIGN

Ocular Findings: Absence of normal wrinkling of the forehead on looking upward.

Associated Clinical Findings: Graves' disease.

Increased basal metabolism.

Tremors.

Tachycardia.

Other systemic signs of hyperthyroidism.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

JOHNSON'S SYNDROME

Synonym: *Adherence Syndrome*.

Similar Conditions: *Duane's Syndrome, Moebius Syndrome, Turk's Syndrome*.

Ocular Findings: Adhesions of the lateral rectus muscle sheath to the inferior oblique resulting in paresis of the lateral rectus muscle and consequent limited abduction.

Adhesion between the superior rectus muscle sheath and the superior oblique with resulting limitation of elevation.

Associated Findings: May have head tilt.

Bibliography

Johnson, L. V.: Adherence syndrome; pseudoparalysis of lateral or superior rectus muscles. *Arch. Ophthalm.*, 44:870-878, 1950.

JOHNSON-STEVEN'S DISEASE

Synonyms: *Stevens-Johnson Syndrome, Hebra's Syndrome*.

Ocular Findings: Purulent pseudomembranous conjunctivitis.
Bulbar chemosis.

Symblepharon.
Keratitis and iritis.
Hypopyon.

Associated Clinical Findings: Erythema multiforme.
Stomatitis.
Rhinitis.
Balanitis and urethritis.
Frequent respiratory infections.

Ethiology: Unknown. May be partly due to drug toxicities or allergies.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Robinson, H. M., Jr. and McCrumb, F. R., Jr.: Comparative analysis of mucocutaneous-ocular syndromes: report of 11 cases and review of literature. *Arch. Dermat. and Syph.*, 61:539-560, 1950.

K

KAPOSÍ'S DISEASE

Synonym: *Xeroderma Pigmentosum*.

Ocular Findings: Pigmented spots or ephelides of lentigo type in the conjunctiva.
Chronic conjunctivitis.
Conjunctival ectropion in severe cases.
Papillomas of the conjunctiva and eyelids.

Associated Clinical Findings: Erythematous and vesicular eruptions on the uncovered parts of the body (face, arms, neck, and hands).
Usually begins in the second or third decades.

Etiology: Familial.

Note: This eponym should not be confused with Kaposi's hemorrhagic sarcoma; a rare tumor sometimes affecting the lids.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

KAYSER-FLEISCHER RING

Ocular Findings: Gray-green to golden-red colored pigmented ring appearing on the posterior surface of the cornea, located in the periphery of Descemet's membrane.

This ring is usually one to three millimeters wide and displays a blurred inner margin and a distinct outer margin.

It is sometimes outlined inside by a second circle.

Associated Clinical Findings: Wilson's Disease (hepatolenticular degeneration).

Pathology: The Kayser-Fleischer ring is felt to be a copper-amino acid compound.

It is actually a ring of microscopic pigmented granules.

Bibliography

Donaldson, D.: Kayser-Fleischer ring in hepatolenticular degeneration (Wilson's disease). *Arch. Ophthalmol.*, 72:116-117, 1964.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

KENNEDY SYNDROME

Synonyms: *Foster-Kennedy Syndrome, Gowers-Paton-Kennedy Syndrome.*

Ocular Findings: Homolateral scotoma (central).

Homolateral retrobulbar neuritis and optic atrophy.

Contralateral papilledema.

Associated Clinical Findings: Anosmia may be present.

Pathologic Etiology: Tumor, abscess or aneurysm in the base of the frontal lobe or sphenoid crest. Prognosis depends on the type of lesion.

Bibliography

Masters, S.: Foster-Kennedy syndrome. *Amer. J. Ophthalmol.*, 36:983-985, 1953.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

KHUNT-JUNIUS DISEASE

Synonym: *Disciform Degeneration of the Macula.*

Ocular Findings: Loss of central vision.

Metamorphopsia.

Positive central scotoma.

Early, the lesion appears as a dark, central elevated retrolental mass without degenerative changes of the retina and pigment epithelium in that area.

The lesion may be difficult to distinguish from a malignant melanoma of the central choroid.

Later, degeneration of the pigment epithelium of the central retina occurs, as well as ingrowth of blood vessels and scar tissue from the choroid, resulting in a disc-shaped yellowish-white mound in the macular area.

Disciform degeneration of the macula is a disease of advanced age and usually involves both eyes.

Pathology: Blood and serum are found between Bruch's membrane and the pigment epithelium. Also, breaks in Bruch's membrane in the foveal choroid may be found.

Bibliography

Hogan, M. J., and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Klien, B.: Diseases of the macula: basic histopathologic processes in retina, pigment epithelium and choroid which modify their clinical appearance. *A.M.A. Arch. Ophthalmol.*, 60:175-186, 1958.

Wolter, R.; Beng, C. A.; and Roth, F. D.: Early disciform degeneration of the macula. *Amer. J. Ophthalmol.*, 59:870-875, 1965.

KILOH-NEVIN SYNDROME

Ocular Findings: Progressive eye muscle pareses.

Bilateral ptosis.

Associated Clinical Findings: Progressive weakness of the facial, neck, and shoulder muscles.

Elevated blood aldolase.

Pathology: May appear as a form of progressive muscular dystrophy with non-nuclear basis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

KIMMELSTIEL-WILSON'S DISEASE

Synonyms: *Kimmelsteil-Wilson Syndrome, Intercapillary Glomerulosclerosis.*

Ocular Findings: This syndrome, which occurs in diabetics, shows a more or less characteristic ophthalmoscopic picture. In addition to the changes characteristic of diabetic retinopathy and hypertensive retinopathy, the veins are found to be enormously engorged.

The fundus takes on the picture of advanced diabetic retinopathy and grade three to four hypertensive retinopathy.

Retinopathy has been reported in over ninety per cent of verified cases of the Kimmelsteil-Wilson syndrome.

Pathologic Etiology: Diabetes mellitus. Control of the diabetes is reflected in the degree of retinopathy.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

KLAUDER'S SYNDROME

Ocular Findings: Hemorrhagic conjunctivitis.

Subconjunctival ecchymoses.

Symblepharon may be a complication.

May have visual loss.

Associated Clinical Findings: Vesicular eruption and inflammation of the mucous membranes and skin of the trunk, face, and extremities.

The nose, mouth, urethra, vagina, and anus may be involved.

Pneumonia is a common complication.

Pathologic Etiology: May be a variant of erythema multiforme.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
- Klauder, J. V.: Ectodermosis erosiva pluriorificialis; its resemblance to human form of foot and mouth disease and its relation to erythema exsudativum multiforme. *Arch. Dermat. and Syph.*, 36:1067-1077, 1937.

KLEIN-WAARDENBURG SYNDROME

Ocular Findings: Lateral displacement of the internal canthi and the lacrimal puncta.

Hyperplasia of the medial portion of the eyebrows.

Heterochromia of the iris.

Associated Clinical Findings: Hyperplasia and enlargement of the root of the nose.

White forelock.

Congenital deafness which may be unilateral or bilateral.

Etiology: Familial, dominant inheritance.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.
- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo. 1961.

KLINEFELTER'S SYNDROME

Ocular Findings: In the normal population, color blindness occurs in eight per cent of males. Color blindness is absent in Klinefelter's.

Associated Clinical Findings: Klinefelter's patients are phenotypically males but their sex chromatin is positive as in females.

Clinically, they have small testes with sterility, increased urinary gonadotropins and often have gynecomastia and eunuchoid physique.

Pathology: The clinical picture is explained by the presence of the two X-chromosomes. Color blindness does not appear in heterozy-

gotes because the effect of the color-blind gene on one X-chromosome is suppressed by the normal gene on the other X-chromosome. This is also what happens in females.

Bibliography

Francois, J.: Chromosome abnormalities and ophthalmology. *J. Pediat. Ophthalm.*, 1:5-18, 1964.

KLIPPEL-FEIL SYNDROME

Synonym: *Congenital Brevicollis and Craniostenosis.*

Ocular Findings: Congenital paralytic strabismus.

May have hypertropia.

May be associated with Duane's syndrome.

Associated Clinical Findings: Short neck with immobility and torticollis.

Webbed neck.

Spina bifida.

Mental deficiency.

Congenital upward displacement of the scapula (Sprengel's deformity).

Etiology: Congenital.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

KLOEPFER'S SYNDROME

Ocular Findings: Complete amaurosis.

Optic atrophy.

Associated Clinical Findings: Mental deficiency.

Infantilism.

Onset in infancy.

Mortality usually by early adulthood.

Etiology: Hereditary. Autosomal recessive.

Bibliography

McKusick, V. A.: *Medical Genetics*. C. V. Mosby, St. Louis, Mo., 1961.

KNIE'S SIGN

Ocular Findings: Unequal dilation of the two pupils in dim illumination after light stimulation.

May be present as a permanent anisocoria.

The difference is slight and is not a frequently seen sign.

Associated Clinical Findings: Graves' disease.

Increased basal metabolism.

Tachycardia.

Other signs of the thyrotoxic state.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

KOBY'S CATARACT

Synonym: *Floriform Cataract*.

Ocular Findings: Many annular and flattened spacities of variable dimensions and colors are arranged around the anterior and posterior sutures of the embryonic nucleus.

The opacities appear bluish to grayish.

Vision is diminished.

Etiology: Congenital.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

KOCHER'S SIGN I

Ocular Findings: Increased retraction of the lids on fixation.

Lasts only briefly.

Is frequently visible in normal individuals.

KOCHER'S SIGN II

Ocular Findings: Globe lag (reverse von Graefe's).

In looking upward, the lids move upward faster than the eye.

An infrequent sign.

Associated Clinical Findings: Graves' Disease.

Increased basal metabolism.

Tachycardia.

Other signs of hyperthyroidism.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

KOEPPE NODULES

Ocular Findings: Translucent nodules on the pupillary border of the iris.

Seen in cases of granulomatous uveitis, such as sarcoidosis.

Pathology: These nodules consist primarily of epithelioid cells.

They are usually evanescent and usually appear in small crops and disintegrate and absorb after a few days. In severe cases they become organized and posterior synechias develop at their sites.

They are indicative of inflammation of the posterior ocular segment or of the anterior choroid and ciliary body.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

KOPLIK'S SPOTS

Ocular Findings: Round or oval pale spots which appear on the conjunctival mucous membranes and caruncle as well as in the mouth in measles.

The spots are surrounded by a faintly inflammatory areola.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

KORNZWEIG-BASSEN SYNDROME

Synonym and Similar Conditions: *Bassen-Kornzweig Syndrome, Refsum's Syndrome.*

Ocular Findings: Retinitis pigmentosa.

Associated Clinical Findings: Spinocerebellar type degeneration.

Celiac disease.

Signs overlapping those of Refsum's syndrome.

Pathology: Absent serum beta-lipoprotein.

A "spiny" appearing erythrocyte (acanthrocyte).

In Bassen-Kornzweig syndrome, the deficiency of beta-lipoprotein lowers all serum lipids, including the fat soluble vitamin A, leading to a degeneration of the visual cells.

Bibliography

Gills, J. P.; Hobson, R.; Hanley, W. B.; *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. *Arch. Ophthal.*, 74:596-603, 1965.

KRABBE'S DISEASE

Synonym: *Acute Infantile Sclerosis.*

Similar Conditions: *Schilder's Disease, see Pelizaeus-Merzbacher Disease.*

Ocular Findings: Nystagmus.

Optic atrophy.

Progresses to blindness.

Associated Clinical Findings: Acute diffuse cerebral sclerosis in early childhood.

Tonic spasms from touch or noise stimulus.

Rapidly fatal.

Pathologic Etiology: A demyelinating disease of childhood with demyelination of the white matter and secondary gliosis described as diffuse sclerosis.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

KRABBE'S SYNDROME

Synonym: *Encephalotrigeminal Angiomatosis*.

Similar Condition: *Sturge-Weber Syndrome*.

Ocular Findings: Choroidal angiomas.

Hydrophthalmia or late simple glaucoma.

May have detachment of the retina.

May have angiomatosis of the conjunctiva and episclera.

Heterochromia iridis.

Associated Clinical Findings: Cutaneous angiomas (nevus flammeus) of trigeminal distribution.

Facial hemihypertrophy.

Etiology: Congenital.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

KRAUPA'S EPITHELIAL DYSTROPHY

Ocular Findings: Appearing congenitally or starting soon after birth, the epithelium acquires a matt appearance due to the existence of very fine vesicles.

The corneal parenchyma may show a more or less diffuse abnormality.

Etiology: This type of dystrophy is inherited as a recessive and is related to the congenital parenchymatous dystrophies of the cornea such as Meesman's and Francois'.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

KRAUSE'S SYNDROME

Synonym: "*Trisomy Syndrome.*"

Ocular Findings: Bilateral retinal dysplasia.

Microphthalmos.

Severe retinal detachments.

Massive retinal hemorrhages.

Ptosis.

Strabismus.

Complications such as secondary cataracts and glaucoma may occur.

Associated Clinical Findings: Mental deficiency.

Hydrocephalus.

Cerebellar hypoplasia.

Etiology: Unknown. Prognosis poor.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Krause, A. C.: Congenital encephalo-ophthalmic dysplasia. *Arch. Ophthal.*, 36:387-444, 1946.

KRIMSKI LIGHT REFLEX

Synonym: *Krimski's Sign.*

Findings: This is a method of estimation of the angle of squint deviation by interposing prisms before the deviating eye (and sometimes the fixing eye as well) to obtain centration of a light reflex in the same location in each cornea in reference to the pupils. The prism compensates for the angle of deviation and is therefore measured in "prism diopters."

KRUKENBERG SPINDLE

Ocular Findings: Uveal pigment deposition on the central endothelium in a vertical spindle shape.

Occurs normally in a small percentage of people over twenty, especially myopic women.

About ninety per cent of cases are bilateral.

They are more marked, more frequent, and occur at an earlier age in pathologic states.

Spindle type deposition of uveal pigment is particularly prominent in cases of pigmentary glaucoma and ocular disease with involvement of uveal tissue.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

KUFS' SYNDROME

Similar Condition: *Tay-Sach's disease. Adult form is known as Mulberger's Disease.*

Ocular Findings: Pigmentary changes in the retina.

May have nystagmus.

May have strabismus.

Visual loss in infancy or early childhood.

Hemeralopia.

Associated Clinical Findings: Felt to be a form of juvenile familial idiocy.

Progressive dementia.

Somnolence.

Muscular pareses.

Occasional epileptic seizures.

Pathology: Lipoid degeneration of the ganglion cells of the retina and CNS.

Etiology: Familial. Simple recessive inheritance.

Bibliography

Proceedings of VIII Argentine Congress Ophth., Mendoza, 1966.

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

L

LAFFER-ASCHER SYNDROME

Similar Condition: *Ascher's Syndrome.*

Ocular Findings: Swollen lids, especially the upper lids (blepharochalasis).

Associated Clinical Findings: Swollen lips.

Etiology: Felt to be due to an endocrine imbalance.

Bibliography

- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Orlowski, W. J.; Stepniak, R.; and Zwierzchowski, R.: Le syndrome de Laffer-Ascher étude clinique et pathogenique. *Ann. Ocul.*, 196:362-387, 1963.

LAGLEYZE-VON HIPPEL DISEASE

Synonyms: *Von Hippel-Lindau Syndrome, Lindau Syndrome, Angiomatosis Retinae.*

Ocular Findings: Angiomas of the retina.

May have hemorrhages or exudates in the angiomatous areas.

Associated Clinical Findings: Calcification in the cerebellum or cerebrum is not uncommon.

May have cysts or tumors in the cerebellum, spinal cord, pancreas, or kidneys.

Etiology: Unknown. Some are felt to be familial.

Bibliography

- Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

LANDRY'S DISEASE

Synonyms: *Acute Ascending Paralysis, Guillain-Barré Syndrome.*

Ocular Findings: External ophthalmoplegia.

Inability to completely close the eyelids because of facial palsy.

May find paralysis of accommodation.
May have dilated fixed pupils.
May have optic neuritis and optic atrophy.

Associated Clinical Findings: Neuritis with flaccid paralysis affecting all four limbs usually following a brief febrile illness.
Associated unilateral or bilateral facial paralysis.
Cerebellar ataxia.
Loss of tendon reflexes.

Etiology: Uncertain. In some cases, a preceding episode of acute infectious process with fever is reported.

Bibliography

Ford, F. R.: *Diseases of the Nervous System in Infancy, Childhood and Adolescence*, 3rd ed. Charles C Thomas, Springfield, Ill., 1952.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

LAUBER'S DISEASE

Synonym: *Fundus Albipunctatus Cum Hemeralopia*.

Ocular Findings: Congenital hemeralopia.
Fundus findings characterized by a gray or whitish mottled appearance.
Visual acuity is normal.
Electroretinographic curve is normal.

Etiology: Familial.

Bibliography

Franceschetti, A. and Chome-Bercioux, N.: Fundus albipunctatus cum hemeralopia (cas stationnaire depuis 49 ans). *Ophthalmologica*, 121:185-193, 1951.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

LAUNOIS' SYNDROME

Synonyms: *Pituitary Giantism Syndrome*, *Lanois* (variant spelling), see *Marie's Syndrome*.

Ocular Findings: Visual field defects (secondary to enlargement of pituitary).

Associated Clinical Findings: Characteristics of primary pituitary gigantism.

Males primarily affected.

Onset usually prior to puberty. If onset is after puberty, it may present as acromegaly.

Clinical features may include striae atrophicae.

May have subnormal mentality.

May have cephalalgia secondary to enlarged pituitary.

Pathology: Enlarged pituitary gland associated with hyperplasia of acidophilic cells or a chromophobe adenoma.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Launois, P. and Roy, P.: *Études biologiques sur le Géants*. Masson, p. 50, 1904.

LAURENCE-MOON-BIEDL SYNDROME

Synonyms: *Bardet-Biedl Syndrome, Laurence-Moon Syndrome.*

Ocular Findings: Retinitis pigmentosa.

Iris coloboma may be seen.

Optic atrophy.

Ring scotomas.

Nyctalopia.

Nystagmus.

Strabismus.

Ptosis.

Epicanthus.

Associated Clinical Findings: Obesity of Frolich's type.

Mental retardation.

Hypogenitalism.

Occurs mostly in males.

Prognosis good.

Pathology: Abiotrophy of neuroepithelium.

Etiology: Hereditary variable. Recessive sex linked.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Macklin, M. T.: The Laurence-Moon-Biedl syndrome; a genetic study. *J. Hered.*, 27:97-104, 1936.

LEBER'S CELLS

Findings: Leber's cells are large macrophages containing necrotic material, found in material expressed from trachoma follicles.

LEBER'S DISEASE

Synonym: *Optic Atrophy-amaurosis-pituitary Syndrome.*

Ocular Findings: Unilateral or bilateral optic neuritis.

Peripheral vision usually retained.

Rapid loss of central vision.

Sheathing of retinal vessels with hemorrhages and exudates.

May have papilledema or optic atrophy.

May get partial recovery.

Associated Clinical Findings: Headaches.

Vertigo.

Occasional signs of disseminated sclerosis.

Usually found in males aged twenty to thirty.

Etiology: Hereditary. Male, sex-linked recessive.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

LEBER'S RETINITIS

Synonym: *Leber's Serofibrinous Exudative Retinitis.*

Similar Condition: *Coats' Disease.*

Ocular Findings: Exudative retinitis.

Clinical appearance of "pseudoglioma," and solid detachment of the retina.

Normal anterior segment.

Occurs primarily in male children. Under age one in more than fifty per cent of cases.

Etiology: Uncertain.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

LEBER'S SYNDROME

Synonyms: *Leber's Congenital Amaurosis, Hereditary Retinal Aplasia.*

Ocular Findings: Congenital amaurosis.

Slight or absent pupillary reactions.

Pendular nystagmus.

Cataracts.

Keratoconus or keratoglobus.

Hemorrhages and edema of the retina.

May have optic atrophy.

Pathology: Dysgenesis of neuroepithelium. May resemble retinitis pigmentosa.

Etiology: Familial. Autosomal recessive.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Gillespie, F. D.: Congenital amaurosis of Leber. *Amer. J. Ophthalm.*, 61:874-880, 1966.

Leber, T.: Ueber retinitis pigmentosa und angeborene amaurose. *Arch. F. Ophthalm.*, 15:1, 1867.

LEIGH'S SYNDROME

Synonym: *Leigh's Encephalomyelopathy*

Ocular Findings: Absence of pupillary reaction to light.

Associated Clinical Findings: Generalized muscle weakness.

Hypotonia or hypertonia.

Difficulties in coordination.

Ataxia.

May have progressive mental deterioration.

Mainly affects young children.

Pathology: Thought to be an inborn error of metabolism.

Respiratory embarrassment due to acute damage of the brain stem often leads to death.

Etiology: Familial.

Bibliography

Kearns, T. P.: Neuro-ophthalmology. Annual Review, Arch. Ophthal., 76:729-755, 1966.

Ebels, E. J.; Blokzijl, P. J.; and Troelstra, J. A.: A Wernicke-like encephalomyelopathy in children (Leigh), an inborn error of metabolism? Report of 5 cases with emphasis on its familial incidence, Helv. Paediat. Acta., 20:310-324, 1965.

LEINER'S DISEASE

Synonym: *Infantile Exfoliative Erythroderma.*

Similar Condition: *Wilson-Brocq Disease.*

Ocular Findings: Exfoliative dermatitis of the lids.

Secondary keratitis, and corneal ulcers.

May be complicated by symblepharon.

Corneal sensitivity is diminished.

Associated Clinical Findings: Generalized exfoliative dermatitis occurring in infancy.

May be congenital.

Secondary atrophy of the skin.

Etiology: Unknown.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

LE JEUNE'S SYNDROME

Synonym: *"Cat-Cry" Syndrome.*

Ocular Findings: Hypertelorism.

Epicanthus.

Oblique palpebral fissures.

Associated Clinical Findings: Mental retardation.
Stunted growth.

Pathologic Etiology: Congenital. Has been ascribed to deletion of a portion of the short arm of chromosome No. 5.

Bibliography

Macintyre, M. N.; Staples, W. I.; LaPolla, J.; and Hempel, J. M.: The "cat-cry" syndrome. *Amer. J. Dis. Child.*, 108:538-542, 1964.

LENOBLE-AUBINEAU SYNDROME

Synonym: *Nystagmus-myoclonia Syndrome.*

Ocular Findings: Congenital nystagmus associated with fasciculations of the muscles spontaneously elicited or produced by cold or mechanical stimulation.

Associated Clinical Findings: Tremors of the head and limbs.
Vasomotor disturbances.
Hyperactive reflexes.

Etiology: Familial.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.
Lenoble, E. and Aubineau, E.: Nystagmus-myoclonia syndrome. *Rev. Med. (Paris)*, 26:471, 1906.

LERI'S DISEASE

Synonym: *Leri's Pleonosteosis.*

Ocular Findings: Stiffness and thickening of the eyelids.
Diffuse clouding of the cornea.

Associated Clinical Findings: Stiffness of the joints.
Overdevelopment of the soft tissues around the joints without any other signs of rheumatoid arthritis.

Pathology: Premature ossification of the bony epiphyses.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

LHERMITTE'S SYNDROME

Synonym: *Anterior Internuclear Ophthalmoplegia Syndrome.*

Ocular Findings: Ocular palsy with nystagmus.

Paralysis of adduction during attempted lateral deviation of the eyes.

Convergence is not affected.

Nystagmus occurs in the eye that is in the *abducted* position, but there is little or none in the eye that shows the *adduction* palsy.

Vertical nystagmus on upward gaze is also present.

Pathologic Etiology: May be the result of multiple sclerosis.

May be associated with vascular disease or tumor of the pons.

Prognosis is poor.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

LIGNAC-FANCONI SYNDROME

Ocular Findings: Needle-like cystine crystals in the corneas, conjunctiva, and sclera.

Associated Clinical Findings: Systemic cystinosis.

Usually apparent in early childhood.

Dwarfism.

Renal glycosuria.

Prognosis poor.

Pathology: Accumulation of cystine crystals in the cells of the reticulo-endothelial system.

Etiology: Familial. Autosomal recessive.

Bibliography

Cogan, D. G.: Cystinosis in an adult. *JAMA*, 164:394-396, 1957.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

LINDAU-VON HIPPEL SYNDROME

Synonym or Similar Condition: *Von Hippel Disease.*

Ocular Findings: Angiomas of the retina.

May be complicated by hemorrhages and retinal detachment.

May be complicated by glaucoma.

May have iris angiomatosis.

Associated Clinical Findings: Cystic angiomas of the cerebellum with consequent cerebellar signs.

Polycystic pancreas and kidneys.

Usually seen in young men.

May have epilepsy.

Etiology: Familial in about twenty per cent of the cases.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Joe, S. and Spencer, W. H.: Von Hippel-Lindau disease. *Arch. Ophthalm.*, 71:508-509, 1964.

LITTLE'S DISEASE

Synonym: *Cerebral Diplegia.*

Ocular Findings: Optic atrophy.

Nystagmus.

Strabismus.

May have congenital cataracts.

May be sensitive to atropine and homatropine.

Associated Clinical Findings: Bilateral hemiplegia, worse in the legs which are flaccid rather than extended.

Progressive mental retardation and convulsive seizures are likely.

Instillation of atropine or homatropine into the conjunctival sac may cause death due to extreme hypersensitivity in some cases.

Death usually results in early childhood from intercurrent infection.

Usually detected in early infancy.

Etiology: Congenital.

Bibliography

Lloyd, Lois A.: (chap) *The Eye In Childhood*. Year Book Medical Publishers, Chicago, Ill., 1967.

LOBSTEIN'S DISEASE

Synonyms: *Osteopsathyrosis, Osteogenesis Imperfecta, Van Der Hoeve's Syndrome.*

Ocular Findings: Blue sclera, of a uniform blue because of its transparency and perhaps partly due to thinning of the tissue.

May have keratoconus.

May have megacornea.

May be complicated by glaucoma.

May have cataracts.

Associated Clinical Findings: Bony fragility resulting in multiple spontaneous fractures.

Deafness appearing in early adulthood due to otosclerosis or laxity of the ear ossicles.

Etiology: Familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Ruedemann, A. D., Jr.: Osteogenesis imperfecta congenita and blue sclerotics; clinico-pathologic study. *A.M.A. Arch. Ophthal.*, 49:6-16, 1953.

LOEWI'S SIGN

Ocular Findings: Rapid dilatation of the pupils by weak concentration of adrenalin.

This is a sign of increased sympathetic tonus.

An infrequent finding.

Associated Clinical Findings: Graves' Disease.

Increased basal metabolism.

Tremors.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

LÖFFLER'S SYNDROME

Synonym: *Eosinophilic Pneumonia Syndrome.*

Ocular Findings: Frequent retinal infarcts (secondary to pulmonary disease).

Associated Clinical Findings: Pulmonary infiltration and pneumonia with malaise, weakness, cough, and pain in the chest.
May have bronchial asthma.

Pathology: Eosinophilia of the blood may reach eighty per cent.

Etiology: Felt to be a variant of the hypersensitivity state.

Bibliography

Rodman, T.; Fraimow, W.; and Mgerson, R. M.: Löffler's syndrome: report of a case associated with administration of mephensin carbamate (tolseram). *Ann. Intern. Med.*, 48:668-674, 1958.

LOUIS-BAR SYNDROME

Synonym: *Oculo-cerebellar Telangiectasia.*

Ocular Findings: Conjunctival telangiectasia.
Clinical appearance like "pink eye."

Associated Clinical Findings: Cerebellar telangiectasia.
Cerebellar ataxia.
May find telangiectasia of the ear lobes.

LOWE'S SYNDROME

Synonym: *Oculocerebrorenal Syndrome.*

Ocular Findings: Nystagmus.
Glaucoma.
Cataracts.
Blue sclera.

Peculiar wart-like excrescences are noted on the lens capsule anteriorly.

Posterior lenticonus may be noted as a result of thinning of the posterior capsule.

Associated Clinical Findings: Mental deficiency.

Systemic acidosis and aciduria.

Renal rickets.

Generalized hypotonia.

Etiology. Congenital.

Bibliography

- Adams, S. T.; Grant, W. M.; and Smith, T. R.: Congenital glaucoma (possibly Lowe's syndrome). *Arch. Ophthal.*, 68:191-195, 1962.
- Johnson, L. V.: Tonographic survey. *Amer. J. Ophthal.*, 61:680-689, 1966.
- Lowe, C. U.; Terry, M.; and MacLacklan, E. A.: Organic-aciduria, decreased renal ammonia production, hydrophthalmos, and mental retardation; clinical entity. *Amer. J. Dis. Child.*, 83:164-184, 1952.

LUTZ'S SYNDROME

Synonym: *Anterior Internuclear Ophthalmoplegia Syndrome.*

Ocular Findings: Bilateral extraocular muscle palsy characterized by paralysis of both internal recti during lateral gaze, with normal convergence.

Occasionally may be unilateral.

The external rectus on the side of the lesion functions normally or is only slightly paretic, while the contralateral internal rectus does not function in conjugate lateral gaze toward the side of the lesion, although convergence is preserved.

Bibliography

- Hyde, J. E. and Slusher, M. A.: Functional role of median longitudinal fasciculus in evoked conjugate ocular deviations in cats. *Amer. J. Physiol.*, 200:919-922, 1961.
- Orlowski, W. J.; Solmski, P.; and Wojtowicz, S.: Bielschowsky-Lutz-Cogan syndrome. *Amer. J. Ophthal.*, 59:416-430, 1965.

M

MAEDER AND DANIS' DEEP FILIFORM DYSTROPHY

Ocular Findings: Reported in association with keratoconus.

Bilateral corneal opacities in the deep layers of the stroma just in front of Descemet's membrane.

The opacities have the shape of twisted corkscrew filaments.

Etiology: Familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

MAGENDIE-HERTWIG SYNDROME

Synonym: *Skew Deviation*.

Ocular Findings: Skew deviation (monocular hypertropia or vertical divergence) that is referable neither to a peripheral neuromuscular lesion nor to a local mechanical factor of the orbit.

May be concomitant or nonconcomitant.

May have diplopia.

Usually manifested as a maintained deviation of one eye above the other in all positions of gaze.

May simulate vertical muscle palsy.

Pathologic Etiology: Posterior fossa lesion. May follow trauma or cerebellar surgery or basilar artery disease.

The eye on the side of the lesion is usually hypotropic.

Bibliography

Smith, J. L.; David, N. J.; and Klintworth, G.: Skew deviation. *Survey Ophthalmol.*, 11:237-238, 1966.

MALHERBE'S DISEASE

Synonyms: *Benign Calcifying Epithelioma, Hair Matrixoma*.

Ocular Findings: Benign epitheliomas of the lids.

Occurs usually in young adults.

More common in females.

Pathology: Benign tumor arising from the hair matrix cells and producing a calcifying epithelioma. It is comprised of variable proportions of necrotic and viable epithelial cells and may simulate an invasive carcinoma but it does not metastasize.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

MARCHESANI'S SYNDROME

Synonyms: *Weil Marchesani Syndrome*, "*Inverted Marfan's*" *Syndrome*.

Ocular Findings: Spherophakia.

Myopia.

Glaucoma.

Dislocated lenses.

Cataracts.

Microcornea.

Associated Clinical Findings: Brachydactyly.

Short stature.

Hearing defects.

Decreased joint flexibility.

Pathology: Systemic mesodermal dystrophy. Defective closure of fetal fissure.

Etiology: Congenital. Hereditary.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Rosenthal, J. W. and Kloepper, H. W.: Spherophakia-brachymorphia syndrome. A.M.A. Arch. Ophthal., 55:28-35, 1956.

Zabriskie, J. and Reisman, M.: Marchesani syndrome. J. Pediat., 52:158-169, 1958.

MARCUS GUNN PHENOMENON

Synonym: *Jaw-Winking Phenomenon*.

Ocular Findings: Congenital unilateral ptosis.

Paresis of the other ocular muscles supplied by the third nerve, especially the superior rectus, may be present.

Homolateral retraction of the upper lid occurs on opening the mouth or moving the jaw to the opposite side.

In some cases the lid movements are connected with the act of sucking or swallowing.

The lid again droops when the jaw is moved to the homolateral side or when the mouth is closed.

Pathologic Etiology: It is felt that some of the third nerve fibers to the levator originate in the fifth nerve nucleus which supplies the jaw muscles. See Gunn.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Wartenberg, R.: Winking-jaw phenomenon. *Arch. Neurol. and Psychial.*, 59:734-753, 1948.

MARFAN'S SYNDROME

Synonyms: *Arachnodactyly, Dolichostenomelia.*

Ocular Findings: Bilateral dislocation of the lenses.

Occasional megalocornea.

Cataracts.

Secondary glaucoma.

Associated Clinical Findings: Elongation of the long bones.

Muscular hypoplasia.

Deficiency of subcutaneous tissue.

Ear deformities.

Asthenic habitus and infantilism.

Dysraphic disturbances such as spina bifida.

Etiology: Congenital. Hereditary.

Bibliography

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Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Von Noorden, G. K. and Schultz, R. O.: A gonioscopic study of the chamber angle in Marfan's syndrome. *Arch. Ophthal.*, 64:929-934, 1960.

MARIE-STRÜMPPELL'S DISEASE

Ocular Findings: Acute anterior non-granulomatous uveitis.

Occasional episcleritis or scleritis.

Associated Clinical Findings: Ankylosing spondylitis.
Occurs predominantly in young males.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

MARIE'S CEREBELLAR ATAXIA

Synonym: *Spinocerebellar Syndrome*.

Ocular Findings: Nystagmus.
Occasional external ophthalmoplegia.

Associated Clinical Findings: Pyramidocerebellar gait (slow and a bit unbalanced).
Static ataxia.
Chorea-like instability.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

MARIN AMAT SYNDROME

Synonym: "*Inverted Marcus Gunn Syndrome*."

Findings: This is a neurologic anomaly in which one eye closes when the patient's mouth is opened widely or forcibly.

There are various opinions about the neuromuscular mechanisms involved. Some writers have suggested a supranuclear origin. Others insist on a concomitant partial facial paresis and refer to the syndrome as a synkinesis between the facial nerve and the motor branch of the trigeminal.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Wartenberg, R.: Inverted Marcus Gunn phenomenon (so-called Marin Amat syndrome). *Arch. Neurol. and Psychial.*, 60:584-596, 1948.

MARINESCO-SJÖGREN SYNDROME

Ocular Findings: Aniridia.

Congenital cataracts.

Associated Clinical Findings: Congenital cerebellar ataxia.

Oligophrenia.

Dwarfism.

Scoliosis.

Genu valgum.

Defects of the digits.

Etiology: Familial. Autosomal recessive. Frequently the product of consanguineous parents.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Gillespie, F. D.: Aniridia, cerebellar ataxia, and oligophrenia in siblings. *Arch. Ophthalm.*, 73:338-341, 1965.

MARKUS' SYNDROME

Synonyms: *Adie's Syndrome*, *Pupillotonia*.

Ocular Findings: Tonic pupil, usually partially dilated.

Most are unilateral.

Most frequent in young women.

Pupil reacts slowly to light and accommodation but reacts readily to miotics and mydriatics. The dilated pupil is constricted with 2.5% mecholyl which has no effect on the normal pupil.

Associated Clinical Findings: Frequently associated with absent or diminished knee and ankle reflexes.

Etiology: Unknown.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

Laties, A.; Laties, A. M.; and Scheie, H. G.: Adie's syndrome: duration of methacholine sensitivity. *Arch. Ophthalm.*, 74:458-459, 1965.

MARIOTTE'S BLIND SPOT

Synonym: *Physiologic blind spot.*

Ocular Findings: The physiologic blind spot. The boundaries lie approximately between twelve and seventeen degrees from fixation in the temporal visual field.

It is noted farther away from central fixation in hyperopia and closer in myopia.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

MARTIN-ALBRIGHT SYNDROME

Synonym: *Polyvisceral Dystrophy.*

Ocular Findings: Blue sclera.
Cataract.

Associated Clinical Findings: Retarded and stunted growth.

Short upper extremities.

Thick skin.

Teeth and nail anomalies.

Debility.

Polydypsia and polyuria.

Pathology: Low blood calcium. High blood phosphate. May portend latent tetany.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

MAURIAC'S SYNDROME

Ocular Findings: Posterior subcapsular cataracts.
Diabetic retinopathy.

Associated Clinical Findings: Juvenile diabetes.

Osteoporosis.

Defective growth.

Hepatomegaly.

Moon-face.

Defective fat metabolism.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

McCUNE-ALBRIGHT'S SYNDROME

Synonym: *Fuller Albright Syndrome.*

Ocular Findings: Unilateral proptosis.

Papilledema.

Visual field defects secondary to optic atrophy.

Associated Clinical Findings: Disseminated melanosis (brown skin patches).

Precocious puberty.

Etiology: Unknown. Possible female sex linkage.

Bibliography

Hibbs, R. E. and Rush, H. P.: Albright's syndrome. Ann. Intern. Med., 37:587-593, 1952.

MEESMAN'S EPITHELIAL DYSTROPHY

Ocular Findings: Appearance in the epithelium of small, whitish-gray points close together and visible only with the slit-lamp.

In reflected light these points look like vesicles or droplets.

The interpalpebral region of the cornea is more severely affected.

Begins in early infancy.

Sensitivity of the cornea may be reduced.

Visual acuity may be good but later some of the superficial scars from keratitis may reduce vision.

Etiology: Familial. Dominant inheritance.

Bibliography

- Kuwabara, T. and Ciccarelli, E. C.: Meesman's corneal dystrophy; a pathological study. Arch. Ophthalm., 71:676-682, 1964.
- Stocker, F. W. and Holt, L. B.: Rare form of hereditary epithelial dystrophy; genetic, clinical and pathologic study. A.M.A. Arch. Ophthalm., 53:536-541, 1955.

MELKERSSON-ROSENTHAL SYNDROME

Ocular Findings: Non-pitting edema of the eyelids (simulates angio-neurotic edema).

Lagophthalmos.

Blepharochalasis.

Occasionally, peripheral corneal opacities near the lower limbus.

Retrobulbar neuritis.

Paresis of the medial recti.

Bilateral recurrent exophthalmos.

Associated Clinical Findings: Peripheral facial nerve palsy.

Non-inflammatory swelling of the lips or face.

Congenital furrowed tongue.

Has been reported in association with megacolon and craniopharyngioma.

Etiology: Unknown. May have irregular dominant inheritance.

Bibliography

- Paton, D.: The Melkersson-Rosenthal syndrome; a case report. Amer. J. Ophthalm., 59:705-709, 1965.

MENDE'S SYNDROME

Similar Condition: *See Waardenburg.*

Ocular Findings: Poliosis of the lashes and brows.

May have albinotic iris.

Associated Clinical Findings: Congenital deafness.

White forelock.

Mongoloid facies.

Short neck.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

MENIERE'S DISEASE

Ocular Findings: Vestibular Nystagmus (amplitude and duration reduced on the diseased side).

Associated Clinical Findings: Tinnitus.

Paroxysms of vertigo.

Gradual diminution of hearing.

Headaches, nausea, and vomiting related to vertigo.

More common in adult males.

Onset usually in the fifth decade of life.

Prognosis good.

Etiology: Unknown. May be seen with lesions of the inner ear with involvement of the labyrinth.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Elia, J. C.: Drug therapy in Meniere's syndrome: double blind study. *Med. Times*, 93:597-602, 1965.

MÉTÉNIER'S SIGN

Ocular Findings: Extreme laxness of the lids, producing easy eversion of the upper eyelids.

This eversion tends to occur by itself if the thin loose skin of the upper lid is pulled upward.

Associated Clinical Findings: Ehler's-Danlos syndrome. (Fibrodysplasia hyperelastica).

Méténier's Sign is pathognomonic of this condition.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

MEYER'S SIGN

Ocular Findings: In morbilli (measles), there is an initial appearance of "glassy eyes" before the onset of the exanthem which lasts for one to three days. Shortly after this a peculiar swelling of the plica semilunaris may manifest itself (Meyer's sign) still during the incubation period and before the rash appears. Thus, Meyer's Sign is a premonitory sign of measles.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

MEYER-SCHWICKERATH-WEYERS SYNDROME

Synonym: *Oculo-dental-digital Dysplasia.*

Ocular Findings: Microphthalmia.

Glaucoma.

Iris anomalies.

Associated Clinical Findings: Broad, high nasal bridge.

Hypotrichia.

Flexion deformities of the fingers.

Dental defects.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

MIKULICZ'S DISEASE

Similar Condition: *Mikulicz's Syndrome.*

Ocular Findings: Narrowed palpebral fissures.

Lacrimal gland enlargement.

Blurring of vision.

Decreasing lacrimal secretion.

Associated Clinical Findings: Moonlike facies.

Enlarged salivary glands.

Dryness of the mouth.

Pathology: Benign lymphocytic inflammation of the seromucous glands.

May be a precursor to malignant lymphoma.

Etiology Note: Some authors consider Mikulicz's Disease and Mikulicz's Syndrome to be essentially the same. Both should suggest the possibility of tuberculous granuloma, lymphoma, sarcoid, mumps, leukemia, or Hodgkins' disease.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

MIKULICZ'S SYNDROME

Similar Condition: *Mikulicz's Disease*.

Ocular Findings: Chronic, simultaneous, bilateral enlargement of the lacrimal glands.

Decreased lacrimal secretions.

S-shaped upper eyelid border characteristic of lacrimal gland enlargement.

Associated Clinical Findings: Simultaneous enlargement of the parotid and submaxillary glands.

Decreased salivary secretions.

Chronic course with frequent recurrences.

Etiology: May be caused by tuberculosis, leukemia, sarcoidosis, Hodgkins disease, or lymphoma.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Robinson, B. E. and Hayne, R. M.: Lymphosarcoma producing Mikulicz's syndrome. *Ill. Med. Jour.*, 126:30-32, 1964.

Robinson, B. E. and Hayne, R. M.: Lymphosarcoma producing Mikulicz's syndrome. *Guildcraft*, 39/12:9-16, 1965.

MILLARD-GUBLER SYNDROME

Synonyms: *Facial Hemiplegia Alternans, Abducens-facial Syndrome.*

Similar Conditions: *See Raymond's Syndrome, Foville's Syndrome.*

Ocular Findings: Diplopia.

Internal strabismus.

Unilateral or bilateral paralysis of the external recti (due to sixth nerve involvement).

Associated Clinical Findings: Seventh nerve palsy with ipsilateral facial paralysis.

Contralateral hemiplegia.

Pathologic Etiology: Lesion in the area of the lower pons, affecting the nuclei of the sixth and seventh nerves and fibers of the pyramidal tract.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

MILLER'S SYNDROME

Synonym: *Oculocerebrorenal Syndrome.*

Ocular Findings: Aniridia.

May have cataracts similar to those seen in Lowe's Syndrome.

Associated Clinical Findings: Wilm's tumor.

Hemihypertrophy.

Mental retardation.

Other systemic congenital abnormalities.

Etiology: Congenital.

Bibliography

Miller, R. W.; Fraumeni, J. F.; and Manning, M. D.: Association of Wilm's tumor with aniridia, hemihypertrophy and other congenital malformations. *New Eng. J. Med.*, 270:922-927, 1964.

Zimmerman, L. E. and Fort, R. L.: Congenital malformations of the eye. JAMA, 196:684-692, 1966.

MILROY'S DISEASE

Ocular Findings: Strabismus.

Ptosis.

May have thickening of the lids.

May have distichiasis.

Etiology: Congenital.

MINKOWSKI-CHAUFFARD SYNDROME

Ocular Findings: Mongoloid eyelid slits.

Microphthalmos.

Heterochromia iridis.

Corneal opacities.

Cataracts.

Associated Clinical Findings: Familial hemolytic icterus.

Splenomegaly.

Anemia.

Ears are prominent.

Dysacucia.

Skull anomalies.

May have skeletal anomalies.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

MITTENDORF DOT

Ocular Findings: Whitish dot on the posterior lens capsule.

Appears as a small whitish or gray circle or dot-like opacity a little below and nasal to the posterior pole of the lens at the point of the lenticular attachment of the fetal hyaloid artery.

Represents the remnant of the hyaloid artery.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

MIZUO'S PHENOMENON

Synonym: *Mizuo's Sign*.

Ocular Findings: Grayish-white discoloration of the retina (characteristic of Oguchi's disease) disappearing when the patient is kept in the dark from two to eight hours. It is replaced by the normal red appearance of the fundus.

Mizuo's phenomenon is a pathognomonic finding in the hereditary night blindness of Oguchi.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

MOEBIUS' DISEASE

Ocular Findings: Diplopia.

Strabismus.

Oculomotor paralysis associated with periodic migraine.

Associated Clinical Findings: Periodic migraine.

Pathology: Third nerve involvement believed to be due to compression between the posterior cerebral and superior cerebellar arteries.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

MOEBIUS' SIGN

Ocular Findings: Inability to hold the eyes in a position of convergence.

The convergence of the eyes is insufficient and the nearpoint of convergence is remote.

This sign may be explained mechanically by the pressure of ex-

ophthalmos on both eyes and is not felt to be definitely pathognomonic for Graves' disease.

Associated Clinical Findings: Hyperthyroidism associated with exophthalmic goiter.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

MOEBIUS' SYNDROME

Similar Conditions: See *Foville's Syndrome*, *Millard-Gubler Syndrome*.

Ocular Findings: Congenital sixth nerve paresis with lateral gaze palsy. Sometimes associated with paresis of the medial recti on lateral gaze movements though convergence is preserved.
Occasional ptosis.
Occasional proptosis.

Associated Clinical Findings: Facial weakness and facial diplegia due to seventh nerve paresis, congenital.
May have deafness.
May be associated with deformities such as webbed fingers or toes, supernummary digits, club foot.

Pathologic Etiology: Aplasia of the motor cerebral nuclei. Defective development in the median longitudinal fasciculus.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Sogg, R. L.: Congenital facial diplegia syndrome of Moebius. *Arch. Ophthalm.*, 65:16-19, 1961.

Stansbury, J. R.: Moebius' syndrome; congenital oculofacial paralysis: case report. *Amer. J. Ophthalm.*, 35:256-261, 1952.

MÖLLER-BARLOW SYNDROME

Synonym: *Infantile Scurvy Syndrome*.

Ocular Findings: Exophthalmos.
Orbital hemorrhages, especially conjunctival.

Associated Clinical Findings: Skin and mucous membrane hemorrhages and hematomas.

Usually appears by end of first year of life.

Bone weakness.

Etiology: C avitaminosis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

MONRAD-KROHN SYNDROME

Synonym: *Paratrigeminal Syndrome*.

Similar Condition: *Raeder's Syndrome*.

Ocular Findings: Miosis.

Ptosis.

Relative enophthalmos (Horner's syndrome).

Associated Clinical Findings: Trigeminal neuralgia.

Pathologic Etiology: Peripheral sympathetic paresis.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

MOOREN'S ULCER

Synonyms: *Rodent Ulcer*, *Serpiginous Ulcer*.

Ocular Findings: Ulceration of the cornea beginning as a yellowish-gray infiltrated excavation near the limbus.

Progresses toward the central cornea with a raised border at the advancing edge.

Rarely involves the deep corneal stroma but may extend to the sclera. May be accompanied by conjunctivitis and episcleritis with much pain.

Frequently bilateral.

Occurs in otherwise healthy, usually elderly, individuals.

Etiology: No microbial agent has been incriminated. Possibly trophic.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

MORGAGNIAN CATARACT

Synonym: *Hypermature Cataract*.

Findings: A special type of hypermature cataract in which the products of disintegration cannot escape due to a thick capsule.

The lens becomes a tense white sac, containing a milky fluid in which the upper border of the nucleus is visible in the pupillary area due to the settling of the nucleus to the bottom of the capsule.

If the nucleus then disintegrates, as well as the cortex, the cataract is then known as a cataracta lactea.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

MORGAGNI'S SYNDROME

Ocular Findings: Bilateral optic atrophy.

Visual field loss.

Associated Clinical Findings: Mental retardation.

Obesity.

Hirsutism.

Lethargy and easy fatiguability.

Vertigo.

Tinnitus.

Onset in the fourth decade.

Prognosis good with treatment.

Pathology: Ocular findings secondary to bony changes (hyperostosis) of the frontal bones.

Etiology: Female dominant sex-linked inheritance.

Bibliography

Falconer, M. A. and Pierard, B. E.: Failing vision caused by bony spike compressing optic nerve within optic canal; report of 2 cases associated with Morgagni's syndrome benefited by operation. *Brit. J. Ophthalm.*, 34:265-281, 1950.

MORQUIO'S DISEASE

Synonyms: *Chondro-osteodystrophy, Mucopolysaccharidosis IV, Brailsford's Syndrome.*

Similar Condition: *Hurler's Syndrome.*

Ocular Findings: Widely spaced orbits.

May find a fine diffuse haze in the corneal stroma bilaterally though some authors describe the cornea as clear.

Vision is not affected.

Associated Clinical Findings: Familial chondro-osteodystrophy with dwarfism.

Large head.

Flexion deformities.

Appearance similar to Hurler's disease except that in Morquio's the corneal signs are not as pronounced and the patients are not mentally defective.

Pathology: A mucopolysaccharidosis with abnormal urine excretion.

Etiology: Familial. Autosomal recessive.

Bibliography

Gills, J. P.; Hobson, R.; Hanley, W. B.; *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. *Arch. Ophthalm.*, 74:596-603, 1965.

Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. *Arch. Ophthalm.*, 74:516-520, 1965.

MORQUIO-ULLRICH SYNDROME

Synonyms: *Mucopolysaccharidosis IV, Ullrich Syndrome.*

Findings: Morquio-Ullrich syndrome is a modification or variation of Morquio's (skeletal changes as in Hurler's, with joint stiffness,

wide mouth, prominent maxilla, short nose, wide set eyes) with definite corneal clouding due to gray punctate opacities more marked in the periphery.

Some authors believe that all Morquio's patients will develop corneal clouding if they survive to adolescence.

Extensive mesodermal chamber angle anomalies have been reported.

Etiology: Familial. Autosomal recessive.

Bibliography

Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal mucopolysaccharide metabolism. *Arch. Ophthalm.*, 74:516-520, 1965.

Von Noorden, G. K.; Zellweger, H.; and Ponseti, I. V.: Ocular findings in Morquio-Ullrich syndrome. *Arch. Ophthalm.*, 64:585-591, 1960.

MUNSON'S SIGN

Synonym: *Munson's Keratoconus Sign.*

Findings: Bowing of the lower lid when the patient looks down. A sign of keratoconus. Best seen if the upper lids are raised when the patient looks down so that the free margin of the lower lid bisects the cornea horizontally. The angular curve assumed by the lid margin is a measure of the corneal curvature.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

MYGIND'S FISTULE SIGN

Findings: In the presence of a fistula in the labyrinth, compression of the air in the external ear canal causes nystagmus to the homolateral side (as on warm water stimulation).

Decrease of the air pressure in the canal by aspiration is followed by nystagmus to the opposite side (as with cold water stimulation). This is called Mygind's fistule sign. This sign is not absolute proof of the presence of a labyrinth fistula, because it is sometimes seen also in other conditions such as hereditary lues (in such case is called "fistule sign without fistule" or Hennebert-Alexander sign).

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

N**NAEGLI'S SYNDROME**

Synonym: *Melanophoric Nevus Syndrome.*

Similar Condition: *Bloch-Sulzberger Syndrome.*

Ocular Findings: Nystagmus.

Strabismus.

Pseudoglioma.

Papillitis and optic atrophy.

Findings similar to Bloch-Sulzberger syndrome but in Naegli's Syndrome the ocular findings are not as pronounced.

Associated Clinical Findings: Pigmentary skin changes.

Dental abnormalities.

Impairment of sweat gland function.

Moderate keratosis palmaris and plantaris.

Bibliography

Franceschetti, A. and Jadassohn, W.: A propos de l'incontinentia pigmenti, délimitation de deux syndromes différents figurant sous le même terme. *Dermatologica* (Basel), 108:1-28, 1954.

NAFFZIGER'S SYNDROME

Synonym: *Scalenus Anticus Syndrome.*

Ocular Findings: Ptosis.

Ipsilateral miosis.

Loss of ciliospinal reflex.

Associated Clinical Findings: Weakness of ipsilateral hand grip.

Reduced muscle tonus of ipsilateral upper extremity.

Pathologic Etiology: Compression of the brachial plexus and subclavian artery by the scalenus anticus muscle.

Bibliography

Collins: *Neurologic Diagnosis*. J. B. Lippincott, Philadelphia, Pa., 1962.

NAGER-DE REYNIER SYNDROME

Synonym: *Dysostosis Mandibularis*.

Similar Condition: *See Franceschetti*.

Ocular Findings: Antimongoloid eye slant.

Associated Clinical Findings: Bilateral mandibular hypoplasia.

Aplasia of the jaw joints.

Bilateral ear anomalies (abnormal pinnae).

Clinical Note: When the condition is unilateral it is referred to as the François-Haustrate syndrome.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

NEGRI-JACOD'S SYNDROME

Synonyms: *Jacod's Syndrome, Petrosphenoidal Syndrome*.

Ocular Findings: Ophthalmoplegia.

Optic atrophy.

Unilateral amaurosis.

Associated Clinical Findings: Trigeminal neuralgia.

Cervical lymphadenopathy in thirty per cent of cases.

Pathologic Etiology: Malignant nasopharyngeal tumor or vascular lesion in the petrosphenoidal area.

Bibliography

Godtfredsen, E.: Ophthalmo-neurological symptoms in malignant nasopharyngeal tumours. *Brit. J. Ophthalm.*, 31:78-100, 1947.

NEWCASTLE DISEASE

Synonym: *Viral Pneumoencephalitis in Fowls.*

Ocular Findings: Acute papillary conjunctivitis.
Secondary infiltration of the corneal epithelium.

Associated Clinical Findings: Preauricular node involvement.
Headaches.
Chills.
Mild meningoencephalitis.
Leucopenia.

Pathology: Cytoplasmic inclusion bodies on epithelial scrapings.

Etiology: Viral.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Lippman, O.: Human conjunctivitis due to Newcastle disease virus of fowls. *Amer. J. Ophthalm.*, 35:1021-1028, 1952.

NICOLAS-FAVRE DISEASE

Synonyms: *Poradenitis, Lymphogranuloma Venereum.*

Ocular Findings: Swelling of the eyelids.
Conjunctivitis.
Keratitis of the upper one-third of the cornea.
Occasional scleritis and iritis.
Peripapillary edema and tortuous blood vessels in the posterior poles.
Vision diminished.

Associated Clinical Findings: Genital lymphogranuloma.
Occurs almost exclusively in Negroes.

Pathology: Ulcerative primary lesion with local bubo, followed by fibrosis and sclerosis.

Etiology: Filterable virus. Transmission is by sexual contact.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

NIEDEN'S SYNDROME

Findings: Combination of multiple generalized telangiectases with the development of cataracts in youth or as a young adult.

Associated Clinical Findings: Early development of aortic sclerosis and organic disease of the heart.

Etiology: Unknown.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

NIEMANN-PICK DISEASE

Synonym: *Phosphatide Lipidosis*.

Similar Condition: *Tay-Sach's Disease*.

Ocular Findings: Cherry red spot at the macula (in about sixty per cent of cases).

Progressive decline in vision.

Associated Clinical Findings: Marked hepatosplenomegaly.

Progressive deafness.

Regression in physical and psychic development.

Neurologic disturbances such as systemic hypotonia or spasticity.

Found mostly in Jewish female infants.

Prognosis poor.

May have brownish pigmentation of the skin.

Pathology: Marked hyperlipemia. Felt to be a catabolic disease process.

Etiology: Probable recessive inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Swikowa, K.: Diagnostic difficulties in Niemann-Pick disease occurring in a young child. *Abs. Excerpta Medica*, 18:532, 1964.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

NONNE-MILROY-MEIGE DISEASE

Synonym: *Chronic Hereditary Lymphedema*.

Ocular Findings: Blepharoptosis.

Yellowish chemotic infiltration of the conjunctiva.

Buphthalmos may be present.

Usually congenital but may appear in later childhood.

Pathology: Defective lacrimal drainage.

Etiology: Congenital.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

NORRIE'S DISEASE

Ocular Findings: Total amaurosis at birth.

Congenital bilateral pseudotumor of the retina.

Total retinal detachment.

Extensive ring-shaped synechias and uveal ectropion.

Associated Clinical Findings: Deafness.

Mental deficiency.

Pathology: Most characteristic feature is severe hypoplasia of the inner retinal layers with secondary changes.

Etiology: Familial. Recessive sex-linked X-chromosomal inheritance through females to males.

Bibliography

Andersen, S. R. and Warburg, M.: Norrie's disease; congenital bilateral pseudotumor of the retina with recessive x-chromosomal inheritance; preliminary report. *Arch. Ophthalmol.*, 66:614-618, 1961.

Norrie, G.: Causes of blindness in children: twenty-five years' experience of Danish Institutes for Blind. *ACTA Ophthal.*, 5:357-386, 1927.

NOTHNAGEL'S SYNDROME

Synonym: *Ophthalmoplegia-cerebellar Ataxia Syndrome.*

Similar Condition: *See Brun's Syndrome.*

Ocular Findings: Bilateral partial third nerve paralysis.

May have ptosis of upper lids.

Gaze paresis most frequent upward combined with some degree of internal or external ophthalmoplegia.

Associated Clinical Findings: Somnolence.

Trunkal ataxia.

Paralysis occurs first, then ataxia (compare with Brun's).

Pathology: Lesion in the superior cerebellar peduncle.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Hiller, F.: The vascular syndromes of the basilar and vertebral arteries and their branches. *J. Nerv. Ment. Dis.*, 116:988-1016, 1952.

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

O

OGUCHI'S DISEASE

Ocular Findings: Congenital static hemeralopia usually characterized by a diffuse yellow or gray coloration of the fundus.

A form of congenital night blindness occurring in Japan.

After two to eight hours in complete darkness, the normal color of the fundus returns in practically all cases (Mizuo's phenomenon).

Central photopic visual acuity and color sense are normal.

Etiology: Familial. Autosomal recessive.

Pathology: A syncytium of cells and pigment between the neuroepithelium and pigment epithelium.

Bibliography

- Carr, R. E. and Gouras, P.: Oguchi's disease. *Arch. Ophthal.*, 73:646-656, 1965.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

OHARA'S DISEASE

Synonym: *Japanese Tularemia*.

Ocular Findings: Intense conjunctivitis, usually monocular, involving the palpebral conjunctiva with small yellow ulcerations scattered over the conjunctiva.

Corneal ulcers may occur.

Associated Clinical Findings: Enlargement of the regional lymph nodes, followed by suppurative ulcerations.

Headaches.

Vomiting, chills, and fever.

Usually runs its course in a few weeks.

Pathologic Etiology: Caused by *Bacterium tularensis*. Harbored by wild rabbits and squirrels and transmitted directly to man by contact or by bite of blood sucking fly or tick. May be transmitted by rubbing the eyes while preparing the animal for eating.

Bibliography

- Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

OPPENHEIM'S DISEASE

Synonym: *Amyotonia Congenita*.

Similar Conditions: *Werdnig-Hoffmann Syndrome*, *Charcot-Marie-Tooth Syndrome*.

Ocular Findings: Progressive ocular muscle pareses.

May have nystagmus.

May develop optic atrophy.

Associated Clinical Findings: Generalized weakness.

Complete lack of tone in the muscles of the extremities, trunk, and neck.

May have facial and tongue pareses.

Condition is present at birth but may be overlooked until the infant is several months of age.

Pathology: Lower motor neuron degeneration with diminution in the number of motor cells in the spinal gray matter.

Etiology: Congenital.

Bibliography

Ford, F. R.: *Diseases of the Nervous System in Infancy, Childhood and Adolescence*, 3rd ed. Charles C Thomas, Springfield, Ill., 1952.

OSLER'S DISEASE

Synonyms: *Hemorrhagic Familial Angiomatosis, Rendu-Osler Disease.*

Ocular Findings: Telangiectasis of the bulbar and palpebral conjunctiva.
Spontaneous hemorrhages of the conjunctiva.
Occasional angiomas of the retina.

Associated Clinical Findings: Telangiectasis of the skin, especially of the face, arms, and fingers.
Telangiectases of the mucous membranes of the nose, mouth, pharynx, digestive tract, and genitourinary tract.

Etiology: Familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

OTA, NEVUS OF

Synonym: "Blue Nevus," *Oculocutaneous Melanosis.*

Ocular Findings: Bluish subepithelial nevus of the globe.

May involve any part of the uvea, and uveal pigment may be present in any ocular tissue.

Malignant change rarely occurs.

Associated Clinical Findings: Frequently associated with "Mongolian spot" of the sacrum.

Pathology: Melanoblasts in the corium beneath the epithelium. Cells are morphologically identical in the globe and in the "Mongolian spots" on the sacrum.

Etiology: Congenital.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

P

PAGE'S SYNDROME

Synonym: *Hypertensive Diencephalic Syndrome.*

Ocular Findings: Excessive lacrimation.
Epiphora.

Associated Clinical Findings: Vasomotor instability.
Hypertension.
Tachycardia.
Salivation.

Pathologic Etiology: Irritation of the sympathetic and parasympathetic centers in the diencephalon.

Bibliography

Page, I. H.: A syndrome simulating diencephalic stimulation occurring in patients with essential hypertension. *Amer. J. Med. Sci.*, 190:9-14, 1935.

PAGET'S DISEASE

Synonym: *Osteitis Deformans.*

Ocular Findings: Exophthalmos.
Strabismus.
Degenerative ring opacities of the cornea.
Optic atrophy.
Choroidal sclerosis.

Exudative and hemorrhagic retinopathy.
Angioid streaks of the retina.

Associated Clinical Findings: Hyperplasia of the bones of the cranium and face which gives the patient a simian look.
Bowlegs and kyphoscoliosis.
Onset usually in young adults.
Generalized arteriosclerosis.

Etiology: Familial. Autosomal incomplete dominant.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

PANCOAST'S SYNDROME

Synonym: *Superior Pulmonary Sulcus Syndrome*.

Ocular Findings: Mild enophthalmos (Horner's Syndrome).
Ptosis.
Miosis.

Associated Clinical Findings: Severe shoulder pain on side of lesion.
Paresthesias and weakness of the arm on the affected side.

Pathologic Etiology: Pulmonary apex tumor; most commonly primary bronchogenic carcinoma, though any tumor in this area may produce the syndrome.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
Pancoast, H. K.: Superior pulmonary sulcus tumor; tumor characterized by pain, Horner's syndrome, destruction of bone and atrophy of hand muscles. JAMA, 99:1391-1396, 1932.

PAPPATACT'S DISEASE

Synonym: *Sandfly Fever*.

Ocular Findings: Conjunctivitis.
Occasional neuroretinitis.

Associated Clinical Findings: Systemic chills and fever.

Condition is rare.

Etiology: A viral disease transmitted by the sandfly *Phlebotomus papatasi*.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

PARINAUD'S SYNDROME

Synonym: *Subthalamic Syndrome*.

Ocular Findings: Vertical conjugate gaze palsy.

Frequent paresis of convergence.

Ptosis may be seen.

Occasionally may see displacement of the pupils.

Bell's phenomenon is retained.

May have diplopia.

Associated Clinical Findings: Vertigo.

Occasionally ataxia.

Pathologic Etiology: Lesion in the anterior quadrigeminal plate or roof of the midbrain, such as a pineal gland tumor.

Bibliography

Bielschowski, A.: Lectures on motor anomalies of eyes; paralysis of conjugate movements of eyes. *A.M.A. Arch. Ophthal.*, 13:569-583, 1935.

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

PARINAUD'S SYNDROME

Synonyms: *Parinaud's Oculoglandular Syndrome*, *Cat Scratch Syndrome*.

Ocular Findings: Conjunctivitis with small opaque gray areas of focal necrosis, usually multiple, occurring on any part of the conjunctiva, but most frequently on the palpebral conjunctiva.

Follicular hypertrophy and edema of the lids.

Transient pseudomembranes.

Corneas are almost never involved.
Usually unilateral.

Associated Clinical Findings: Cervical lymphadenopathy.

Fever.

Incubation period three to seven days.

Frequent history of cat scratch or bite.

Prognosis good for recovery in weeks or months.

Etiology: Infection from leptothrix, tuberculosis, or tularemia.

Bibliography

Bedrossian, E. H.: *The Eye*, Charles C Thomas, Springfield, Ill., 1958.

Cassady, J. V. and Culbertson, C. S.: Cat-scratch disease and Parinaud's oculoglandular syndrome. *A.M.A. Arch. Ophthal.*, 50:68-74, 1935.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

PARKINSON'S DISEASE

Ocular Findings: Nystagmus.

Tremor of the eyelids.

Paralysis of convergence.

Occasional accommodative disturbances.

Mydriasis or anisocoria.

Rigidity of eye movements (though the patient can look fully to either side, the eye movements are jerky).

Associated Clinical Findings: Rhythmical tremors.

Sluggishness.

Loss of facial expression.

Seen in late stages of epidemic encephalitis.

May accompany generalized arteriosclerosis.

May be seen in carbon monoxide poisoning.

Pathology: Widespread destruction of pigmented cells of the substantia nigra.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Loeffler, J. D.; Slatt, B.; and Hoyt, W. F.: Motor abnormalities of the eyelids in Parkinson's disease. 76:178-185, 1966.

PARRY-ROMBERG SYNDROME

Synonym: *Facial Hemiatrophy Syndrome.*

Ocular Findings: Horner's Syndrome on the affected side.
Orbicularis paresis has been occasionally reported.

Associated Clinical Findings: Progressive facial hemiatrophy, with atrophy of subcutaneous fat and bone. The muscles are usually not affected.

Migraine headaches are frequently reported.

May have atrophy of the ipsilateral cerebral hemisphere.

Pathologic Etiology: A heredo-degenerative condition.

Bibliography

Mussinelli, F.; Magli, R.; Origlio, A.; and Reti, F.: Progressive facial hemiatrophy. Abs. Amer. J. Ophthal., 60:755, 1966.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

PARRY'S DISEASE

Synonyms: *Exophthalmic Goiter, Thyrotoxicosis, Basedow's Disease, Graves' Disease.*

Ocular Findings: Exophthalmos (Parry's Sign) and any of a number of associated eye and eyelid signs (see under Graves' Disease).

Associated Clinical Findings: Nervousness.

Weakness.

Sensitivity to heat.

Sweating.

Tremors.

Palpitation.

Etiology: Hyperthyroidism. Underlying cause is unknown.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

PARRY'S SIGN

Ocular Findings: Exophthalmos.

Frequently accompanied by other eye signs of thyrotoxicosis.

Associated Clinical Findings: Graves' Disease.

Increased basal metabolism.

Tachycardia.

Occasional tremors and other signs of thyroid dysfunction.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

PASCHEFF'S CONJUNCTIVITIS

Synonym: *Necrotic Infectious Conjunctivitis*.

Ocular Findings: A peculiar type of conjunctivitis characterized by the formation of foci of suppurative necrosing inflammation.

Associated Clinical Findings: Swelling of the regional lymph glands.

Etiology: Microbial; possibly tularemia.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

PASSOW'S SYNDROME

Ocular Findings: Heterochromia iridis.

Cyclitis.

Horner's Syndrome.

Fifth and sixth nerve pareses.

Associated Clinical Findings: Facial hemiatrophy.

Seventh nerve paresis.

Kyphosis.

Funnel chest.

Spina bifida.

Anomalies of the fingers.

Trophic and vasomotor defects of the lower extremities.

Etiology: Felt to be a form of Bremer's status dysraphicus with defective closure of the spinal canal.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

PATAU'S SYNDROME

Synonyms: "*D Trisomy Syndrome*," "*13-15 Trisomy Syndrome*."

Ocular Findings: Microphthalmos.

Anophthalmos.

Retinal dysplasia.

Hypertelorism.

Associated Clinical Findings: Multiple hereditary congenital deformities.

Mental deficiency.

Cleft palate.

Polydactyly.

Horizontal hand palm creases.

Usually fatal in early infancy.

Etiology: Hereditary.

Bibliography

Cogan, D. G. and Kuwabara, T.: Ocular pathology of the 13-15 Trisomy syndrome. Arch. Ophthal., 72:246-253, 1964.

Ginsberg, J. and Perrin, E. V.: Ocular manifestations of 13-15 Trisomy: report of a case with clinical, cytogenetic, and pathologic findings. Arch. Ophthal., 74:487-495, 1965.

PATON'S SIGN

Ocular Findings: Comma-shaped dilations of the lower bulbar conjunctival vessels with sludging of the blood. Seen best under the slit-lamp.

Associated Clinical Findings: Sickle-cell disease. Paton's sign is one of the pathognomonic findings of this condition.

PEL'S SYNDROME

Synonym: *Tabetic Eye Crisis.*

Ocular Findings: Paroxysmal attacks of ciliary neuralgia with corneal hyperesthesia and photophobia.

Tearing.

Lid spasm.

Glaucoma may be found.

Tic secondary to trigeminal irritation.

Associated Clinical Findings: Syphilis.

May have multiple neuralgias.

Etiology: Syphilis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

PELIZAEUS-MERZBACHER DISEASE

Similar Condition: *Krabbe's Diffuse Sclerosis.*

Ocular Findings: Nystagmus early in the disease.

Optic atrophy is frequently seen in later stages.

Associated Clinical Findings: Tremors of the head and hands.

Muscular pareses.

Hyperreflexia.

Progressive mental deterioration.

Appears in infancy but progress of disease is slow and patients may have remissions lasting many years.

Pathology: Similar to Krabbe's diffuse sclerosis associated with involvement of the gray substance but with conspicuous sparing of perivascular areas.

Etiology: Familial. Recessive.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

PENFIELD'S SYNDROME

Synonym: *Autonomic Diencephalic Epilepsy Syndrome.*

Ocular Findings: Periods of lacrimation.

Dilation or contraction of the pupils.

Occasional protrusion of the eyes.

Associated Clinical Findings: Sudden vasodilation of the skin supplied by the cervical sympathetic nerves.

Sudden rise in blood pressure.

Increased pulse rate.

Excitability.

These episodes gradually resolve after minutes to hours.

Pathologic Etiology: These symptoms are characteristic of a disturbance of hypothalamic function and are considered to be due to a focal epileptic discharge in the region of the dorsal nucleus of the thalamus on one or both sides.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Penfield, W.: Diencephalic autonomic epilepsy. *Arch. Neurol. and Psychiat.*, 22:358-374, 1929.

PETERS' ANOMALY

Ocular Findings: Congenital leucoma of the corneal stroma, associated with absence of Descemet's membrane and endothelium in the affected areas.

May have anterior synechias.

May be associated with anterior polar cataract.

Etiology: Congenital. Hereditary.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

PEUTZ-TOURAINÉ SYNDROME

Ocular Findings: Freckles or café-au-lait spots on the eyelids and conjunctiva.

Associated Clinical Findings: Café-au-lait spots on the face, especially near the nostrils and lips.

Occasional involvement of the buccal mucosa and anorectal mucosa.

Intestinal polyposis, especially rectocolic.

Becomes apparent in infancy.

Etiology: Regular dominant inheritance.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

PFAUNDLER-HURLER SYNDROME

Synonyms: *Hurler Syndrome*, *Gargoylism*, *Lipochondrodystrophy*, *Mucopolysaccharidosis I*.

Ocular Findings: Bilateral cloudiness of the corneas, present at birth or in early childhood.

Conjunctiva is thick and inelastic.

May have buphthalmos.

May have megacornea.

May have colobomas.

May have pigmentary retinopathy.

Associated Clinical Findings: Disproportionate dwarfism with skeletal abnormalities.

Large head.

Thick lips and tongue.

Mental retardation.

Sexual infantilism.

Pathology: Urine contains excessive mucopolysaccharides. A disturbance of carbohydrate metabolism.

Etiology: Autosomal recessive inheritance.

Bibliography

- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
- Gills, J. P.; Hobson, R.; Hanley, W. B.; *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. *Arch. Ophthalm.*, 74:596-603, 1965.
- Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharidoses metabolism. *Arch. Ophthalm.*, 74:516-520, 1965.

PICK'S DISEASE

Synonyms: *Arnold-Pick Syndrome, Aphasia-agnosia-apraxia Syndrome.*

Ocular Findings: Apperceptive blindness.

Visual field defects are sometimes present.

Optic neuritis and fixed pupils have been described.

Associated Clinical Findings: Progressive mental deterioration.

Difficulty of speech (motor aphasia).

Occasionally epileptiform seizures.

Occurs more in females from age forty to seventy.

Usually fatal from two to twelve years after onset.

Pathology: Widespread lesion involving occipital lobe. Electroencephalography demonstrates cerebral atrophy.

Etiology: Unknown.

Bibliography

- Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.
- Hassin, G. B. and Levitin, D.: Pick's disease; clinicopathologic study and report of case. *Arch. Neurol. and Psychiat.*, 45:814-833, 1941.
- Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

PICK'S SIGN

Findings: In Sandfly Fever or Pappataci Fever, there is prominent conjunctival injection, limited to the exposed portion of the bulbar conjunctiva (Pick's Sign).

Associated with sudden febrile onset, burning sensations of pain and tenderness in the eyes, accompanied by considerable lacrimation and photophobia.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

PIERRE ROBIN SYNDROME

Ocular Findings: Bilateral congenital high myopia.

Congenital glaucoma.

Retinal disinsertion.

Microphthalmia.

Associated Clinical Findings: Micrognathia (hypoplasia of the mandible).

Glossoptosis.

Cleft palate.

Acute dyspnea.

Episodes of cyanosis.

Dysphagia.

Flattening of the base of the nose.

"Bird-like" appearance.

Etiology: Congenital.

Bibliography

Keitel, H.: Peculiar versus diagnosis facies in pediatrics. *Med. Sci.*, 17/6:47-58, 1966.

Robin, P.: Glossoptosis due to atresia and hypotrophy of mandible. *Amer. J. Dis. Child.*, 48:541-547, 1934.

Smith, J. L.; Cavanaugh, J. J.; and Stowe, F. C.: Ocular manifestations of the Pierre Robin syndrome. *A.M.A. Arch. Ophthalm.*, 63:984-992, 1960.

PILLAT'S PARENCHYMATOUS DYSTROPHY

Ocular Findings: Corneal stromal dystrophy characterized by punctate opacities of various sizes in the middle and deep stroma with a blue-gray appearance.

Between the opacities the stroma is clear.

Numerous small (0.75 mm) glass-like lines may be seen in the peripheral stroma.

Visual acuity and corneal sensitivity are not notably altered.

Etiology: Familial.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

PIPER'S SYNDROME

Ocular Findings: High astigmatism.
Corneal dysplasia.

Associated Clinical Findings: Growth retardation and short stature.
Nail dysplasia with flattened and scoop-shaped fingernails.
Bronchiectasis is occasionally seen.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

PLUMMER-VINSON SYNDROME

Synonyms: *Paterson's Syndrome, Sideropenic Dysphagia Syndrome.*

Ocular Findings: Fissures in the corners of the eyes.
Blepharoconjunctivitis.
Occasional keratitis.
Occasional hemeralopia.

Associated Clinical Findings: Dysphagia.
Stomatitis, glossitis and fissuring of the corners of the mouth.
Atrophic changes in the buccal, glossopharyngeal and esophageal mucous membranes.
Achlorhydria.
May have splenomegaly.
Spoon shaped brittle finger nails.
Commonly in middle aged women.

Pathology: Chronic microcytic hypochromic anemia. Low serum hemoglobin. Mentioned in the literature is the unusual tendency for Plummer-Vinson patients to develop oropharyngeal cancer.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
- von Grósz, S.: Eponym-Syndromes. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

POSNER-SCHLOSSMANN SYNDROME

Synonym: *Glaucomatocyclitic Crisis*.

Ocular Findings: Recurrent unilateral attacks of glaucoma and cyclitis.

The eye does not demonstrate notable ciliary injection.

The pupil may be enlarged.

Vision is only slightly blurred.

The intraocular tension is extremely high.

There is little or no pain associated with the high tension and the eye is not uncomfortable.

KPs may be present at onset and last several days and resolve spontaneously.

Intraocular tension is high for several weeks and returns to normal spontaneously.

Glaucoma tests during symptom-free intervals are negative.

Episodes recur with varying frequency, without residual defects or loss of vision, except after many recurrences.

Condition is self limited.

Etiology: May be a form of autoallergic phenomenon.

Bibliography

- Bouchel, J.; Gerhard, J. P.; and Fétique, P.: New observations on the Posner-Schlossmann syndrome. *Survey Ophthalm.*, 10:379-380, 1965.
- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
- Posner, A. and Schlossmann, A.: Syndrome of unilateral attacks of glaucoma with cyclitic symptoms. *Arch. Ophthalm.*, 39:517-535, 1948.

POTTER'S SYNDROME

Synonym: *Renal Agenesis Syndrome*.

Ocular Findings: Prominent epicanthal folds which form a wide semicircle on each side of the nose.

Hypertelorism.

The line of the palpebral fissures is usually flat or is higher at the outer than at the inner canthus.

Associated Clinical Findings: Prominent crease below the lower lip.

Recession of the chin.

Flat, large, low-set ears.

The nose is slightly flat at the tip.

Etiology: Familial.

Bibliography

Keitel, H.: Peculiar versus diagnostic facies in pediatrics. *Med. Sci.*, 17/6:47-58, 1966.

Passage, E. and Sutherland, J. M.: Potter's syndrome. *Amer. J. Dis. Child.*, 109:80-84, 1965.

PSEUDO-GRAEFE SIGN

Synonym: *Pseudo-Graefe Phenomenon.*

Ocular Findings: Elevation or retraction of the upper lid on looking down, or failure of the lid to follow the eyeball downward even after a lag.

When the eyes are in the primary position, the upper lids may be ptotic.

Pathologic Etiology: Aberrant regeneration of the third nerve. May occur following exophthalmic goiter, trauma, tabes, tumor pressure, anterior poliomyelitis, or vascular lesions of the brain stem.

Bibliography

Bender, M. B.: Nerve supply to orbicularis muscle and physiology of movements of upper eyelid, with particular reference to pseudo-Graefe phenomenon. *A.M.A. Arch. Ophthal.*, 15:21-30, 1936.

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

PURKINJE AFTER IMAGES

Synonyms: *Bidwell's Ghost, Negative After-images.*

Findings: Negative after-images, produced by a modified second stimulus. They are called induced or secondary after-images. They have the opposite qualities as the original image.

For example, if one looks at a white light, the negative after-image (Purkinje after image) will look dark or black.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

PURTSCHER'S DISEASE

Synonyms: *Traumatic Retinal Angiopathy, Purtscher's Retinopathy.*

Ocular Findings: Bilateral edema and exudative retinopathy of the macula and retina secondary to systemic trauma. The vessels may be occluded by fat embolism or air with white exudates between the disc and the macula. Flame-shaped hemorrhages also may be seen.

Vision is diminished.

Usually clears without any permanent damage.

Some writers describe it as a distention of the retinal vessels and arteriolar spasm secondary to fat embolism following crushing injuries of the extremities with fractures of the long bones or crushing injuries of the chest with rib fractures.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Q

QUINCKE'S DISEASE

Synonyms: *Quincke's Edema, Angioneurotic Edema of the Lids.*

Ocular Findings: Sudden painless swelling of the eyelids often preceded by a sensation of fullness or itching.

May be unilateral or bilateral.

May have reduced vision due to corneal edema.

May have transitory myopia.

May disappear spontaneously in a few hours or may last for weeks.

Associated Clinical Findings: Edema may extend into the lower parts of the face.

May be associated with brachial plexus palsy.

Pathologic Etiology: Anaphylaxis, serum sickness, angioneurotic edema.

Bibliography

- Appelbaum, A.: Allergic phenomena in ophthalmology. Arch. Ophthal., 24:803-823, 1940.
Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
Heinrich, Irenaeus Quincke (1842-1922) Clinician of Kiel. JAMA, 196:1152-1153, 1966.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

R

RAEDER'S SYNDROME

Synonym: *Paratrigeminal Syndrome*.

Ocular Findings: Horner's syndrome without loss of the facial sweating mechanism.

Mild enophthalmos, ptosis, unilateral miosis.

May have epiphora.

Occasional diplopia.

Associated Clinical Findings: Facial sweating mechanism is not affected.

May have head and facial pain.

May have weakness of the jaw muscles.

Pathologic Etiology: Lesion in the sympathetic nerve peripheral to the superior cervical ganglion (around the carotid artery) with involvement of the fifth nerve. The facial supply comes off below, hence the lack of involvement of the facial sweating mechanism. Meningiomas and aneurysms of the internal carotid artery are the most frequent causes.

Bibliography

- Bedrossian, E. H.: Raeder's syndrome. A.M.A. Arch. Ophthal., 48:620-623, 1952.
Boniuk, M. and Schlezinger, N. S.: Raeder's paratrigeminal syndrome. Amer. J. Ophthal., 54:1074-1084, 1962.
Raeder, J. G.: Paratrigeminal paralysis of oculo-pupillary sympathetic. Brain, 47:149-158, 1929.

RAMSAY-HUNT SYNDROME

Findings: Paralysis of the facial nerve.

Bell's palsy.

Herpes Zoster involving the inner ear.

RAYMOND'S SYNDROME

Synonyms: *Raymond-Cestan Syndrome, Cestan's Syndrome.*

Ocular Findings: Ipsilateral abducens palsy.
Paralysis of lateral conjugate gaze.

Associated Clinical Findings: Contralateral hemiplegia.
Anesthesia of the face, extremities, and trunk on the involved side.

Pathologic Etiology: Intramedullary lesion of the pons.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

REDLICH'S SIGN

Ocular Findings: Dilation of the pupil on strong muscular effort involving the extremities, such as a strong hand grip.

Associated Clinical Findings: Dementia praecox. Redlich's sign is characteristic of certain forms of dementia.

Pathology: This reaction is apparently an increased psychosensory reflex.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

REESE'S SYNDROME

Synonym: *Reese's Retinal Dysplasia Syndrome.*

Ocular Findings: Bilateral microphthalmia.
Eyes characteristically exhibit severe retinal dysplasia.

Associated Clinical Findings: Polydactylia.
Harelip and cleft palate.
Malformations of the brain, heart, and other viscera severe enough to cause early death in affected infants.

Pathology: Felt to be a form of 13-15 trisomy.

Etiology: Congenital.

Bibliography

Reese, A. B. and Blodi, F. C.: Retinal dysplasia. *Amer. J. Ophthalm.*, 33:23-32, 1950.
Zimmerman, L. E. and Fort, R. L.: Congenital malformations of the eye. *JAMA*, 196:684-692, 1966.

REFSUM'S SYNDROME

Synonym: *Heredopathia Atactica Polyneuritiformis*.

Ocular Findings: Atypical retinitis pigmentosa.

Nyctalopia or hemeralopia.

Constriction of the visual fields.

Pupillary reaction anomalies.

May have cataracts.

May have ptosis.

May have corneal opacities.

Associated Clinical Findings: Chronic polyneuritis.

Progressive paresis in the distal extremities.

Deafness.

Ataxia.

Nystagmus.

Skeletal anomalies.

Pathology: Elevated cerebrospinal fluid protein level without pleocytosis.

Etiology: Familial. Autosomal recessive.

Bibliography

Baum, J. L.; Tannenbaum, M.; and Kolodny, E. H.: Refsum's syndrome with corneal involvement. *Amer. J. Ophthalm.*, 60:699-708, 1965.
Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

REITER'S SYNDROME

Synonym: *Fiessinger-Leroy Syndrome*.

Ocular Findings: Persistent and recurrent inflammation of the bulbar and palpebral conjunctiva with thin mucoid secretion showing few cells.

May have anterior uveitis.

Occasionally may have keratitis.

Associated Clinical Findings: Arthritis.

Urethritis.

Stomatitis and aphthous ulcers of the mouth.

Genital ulcers.

Erythema of the skin.

Primarily affects young men in second to fourth decade.

Etiology: Unknown. Possibly viral.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Foxworthy, D. T.: Adrenocorticotropin and cortisone in treatment of severe Reiter's syndrome. *Ann. Intern. Med.*, 44:52-62, 1956.

RENDU-OSLER-WEBER DISEASE

Synonyms: *Rendu-Osler's Disease, Hereditary Hemorrhagic Angiomatosis.*

Ocular Findings: Telangiectasis in palpebral and bulbar conjunctiva.
Occasional angiomas of the retina.

Associated Clinical Findings: Telangiectases of the skin and mucous membranes.

The cutaneous telangiectases assume a stellar shape with terminal branches.

Conjunctival telangiectases resemble a daisy, the petals of which are formed by capillary arches, tightly placed side-by-side.

Etiology: Hereditary. Regular dominant.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

RIDDOCH'S SYNDROME

Synonym: *Visual Disorientation Syndrome.*

Findings: Visual disorientation in homonymous half-fields without loss of stereoscopic vision. The patients are able to localize objects seen to one side only.

The defect gradually increases from the center towards the periphery of the visual field.

The patient is unaware of the defect as there is little or no disturbance of the central vision.

Loss of visual attention may be present.

Pathologic Etiology: May result from a unilateral lesion of the parietal lobe.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

RIEGER'S ANOMALY

Synonyms: *Rieger's Syndrome, Mesodermal Dysgenesis of the Cornea and Iris.*

Ocular Findings: Appears as an accentuation of a posterior embryotoxon.

A constant feature is the presence of large pectinate-like fibers spanning the irido-corneal angle to be inserted into the periphery of the cornea.

May also have corectopia and colobomas of the iris.

May have buphthalmos.

Infantile glaucoma is not an uncommon sequel.

Associated Clinical Findings: Dental anomalies.

Occasionally dysplasias of the skull, spine, and extremities.

Etiology: Hereditary.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Ernyei, S.: Mesodermal dysgenesis of cornea and iris (Rieger) associated with hypoplasia of the stroma of the iris. *Amer. J. Ophthalm.*, 59:106-108, 1965.

RIFKIND'S SIGN

Ocular Findings: Opaque ring (clinically identical with arcus senilis) at the edge of the cornea.

Pathology: Increased serum fatty acid levels.

Significance: Signifies possible impending heart attack.

RILEY-DAY SYNDROME

Synonym: *Familial Dysautonomia.*

Ocular Findings: Corneal hypesthesia.

Indolent central trophic ulcers.

Decreased tear secretion.

Associated Clinical Findings: Increased salivation with drooling.

Dysphagia.

Increased sweating.

Poor temperature control.

Indifference to pain.

Erythematous blotching of the skin.

Diminished reflexes and muscular hypotonia.

Manifest usually by age two.

Etiology: Hereditary.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

ROBLE'S DISEASE

Synonyms: *River Disease, Onchocerciasis, Guatemalan Disease.*

Ocular Findings: Blindness due to the actual presence of the microfilariae in the eye in all ocular tissues.

Associated Clinical Findings: Systemic cutaneous and subcutaneous infestation with associated symptoms.

Pathologic Etiology: Infestation with the thread worm *Onchocerca*.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

ROCHON-DUVINAUD SYNDROME

Synonym: *Superior Orbital Fissure Syndrome.*

Ocular Findings: Paresis of the oculomotor (third) nerve, superior and inferior branches.

Paresis of the trochlear (fourth) nerve.

Paresis of the abducens (sixth) nerve.

Paresis of the first division of the trigeminal (fifth) nerve, involving the frontal, lacrimal, and nasociliary branches.

Involvement of the optic nerve.

May produce exophthalmos.

Pathology: Lesion involving the superior orbital fissure and producing a sensorimotor ophthalmoplegia. Since the sixth nerve runs below and, later, even between the superior and the inferior branch of the third nerve, presence of a complete third nerve palsy without a sixth nerve palsy excludes the superior orbital fissure as the seat of the lesion.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

ROLLET'S SYNDROME

Synonym: *Orbital Apex-sphenoidal Syndrome.*

Ocular Findings: Varying degrees of exophthalmos.

Ptosis.

Hyperesthesia or anesthesia of the upper lid.

Optic neuritis, papilledema, and optic atrophy.

Visual field defects dependent on optic nerve involvement.

Corneal anesthesia.

Trigeminal neuralgia.

Associated Clinical Findings: Hyperesthesia or anesthesia of the forehead.

Local vasomotor disturbances.

Pathology: Neoplastic, hemorrhagic, or inflammatory lesions in the apex of the orbit.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
- Holt, H. and de Rotth, A.: Orbital apex and sphenoid fissure syndrome. *Arch. Ophthalm.*, 24:731-741, 1940.

ROMAÑA'S SIGN

Findings: Unilateral edema of the eyelids and conjunctiva.

Seen in acute stages of Chaga's disease (trypanosomiasis cruzi).

Primarily seen in children.

Usually seen after about two weeks of fever and signifies a worsening of the disease.

Bibliography

- Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

ROMBERG'S SYNDROME

Synonym: *Progressive Facial Hemiatrophy Syndrome*.

Ocular Findings: Horner's syndrome.

Anterior uveitis.

Heterochromia.

Ptosis and occasionally strabismus.

Ectropion may be seen.

May have trichiasis.

Occasional optic atrophy reported.

Associated Clinical Findings: Progressive atrophy of the skin, muscles, and bones of the face.

Usually unilateral.

Usually starts after puberty.

May have epilepsy.

May have ataxia.

May have systemic cutaneous and skeletal defects.

Etiology: Hereditary. Recessive.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.
- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

ROSENBACH'S SIGN

Ocular Findings: Trembling of the eyelids on gentle closure.

This sign is often found in Graves' disease but is not pathognomonic for it is occasionally seen in other "nervous" persons.

Associated Clinical Findings: Graves' disease.

Tachycardia.

Increased basal metabolic rate.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

ROTH SPOTS

Findings: White spots in the center of retinal hemorrhages.

These spots are characteristic of the retinal hemorrhages of subacute bacterial endocarditis, pernicious anemia, and leukemia.

Roth spots occur particularly if the red blood cells have dropped to below thirty per cent of the normal values.

Such hemorrhages are usually superficial and found scattered around the posterior pole. They are primarily venous and the retinal arterioles maintain their normal appearance.

The cause of the type hemorrhage accompanied by Roth spot is most likely a diminished oxygen supply. As a result, a diapedesis of cells occurs in the center with capillary dilation at the periphery.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

ROTH-BIELSCHOWSKY SIGN

Synonym: *The "Doll Head" Sign.*

Ocular Findings: In rotation of the head to one side or the other, the eyes turn reflexly to the opposite side.

Significance: Seen in paralyzes of voluntary gaze to either side, with intact vestibulo-ocular functions.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

ROTH-BIELSCHOWSKY SYNDROME

Synonym: *Pseudo Ophthalmoplegia.*

Findings: An abnormal type of vestibular nystagmus in which there is an absence of the fast phase of the nystagmus, together with paralysis of voluntary conjugate gaze in one direction. The slow phase is retained.

Stimulation of one labyrinth by irrigation of the ear or rotation of the head causes the eyes to turn reflexly in the direction opposite that of the paralysis.

Stimulation of the opposite labyrinth results in a full deviation of the eyes toward the side of the paralysis.

Pathologic Etiology: Supranuclear (cerebral) lesions, particularly of the temporal lobe.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

ROTHMUND'S DISEASE

Synonyms: *Rothmund's Syndrome, Poikiloderma Atrophicans Vasculare.*

Ocular Findings: Bilateral congenital cataracts.

Cataracts are star-shaped and posterior polar.

Associated Clinical Findings: Atrophic, tight skin with mottling.

Cutaneous telangiectases.

Atrophy of the hair follicles and resultant alopecia.

Ungual dystrophy.

Saddle nose.

Hypogenitalism.

Etiology: Hereditary. Autosomal recessive.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

ROTHMUND-THOMSON SYNDROME

Synonym: *Hereditary Ectodermal Dysplasia.*

Ocular Findings: Juvenile cataracts.

Differs from the classical description of Rothmund's in that cataracts are not necessarily present, and if present, develop later.

Associated Clinical Findings: Ectodermal dysplasia; poikiloderma.

Sparseness of body hair.

Short stature.

Hypogonadism.

Etiology: Hereditary. Autosomal recessive. Rare.

Bibliography

Taylor, W. B.: Rothmund's syndrome; Thomson's syndrome; congenital poikiloderma with or without juvenile cataracts. *A.M.A. Arch. Derm.*, 75:236-244, 1957.

Wahl, J. W. and Ellis, P. P.: Rothmund-Thomson syndrome. *Amer. J. Ophthal.*, 60:722-726, 1965.

ROY'S SYNDROME

Synonym: *Pachydermo-periostosis.*

Ocular Findings: Hypertrophic eyelids, with thickening.

Ptosis.

Madarosis.

Conjunctivitis.

Associated Clinical Findings: Thick hands.

Gyrate skin lines.

Bony abnormalities of the extremities.

Thickening of the skin.

Etiology: An endocrine imbalance is felt to be responsible.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

RUBENSTEIN-TAYBI SYNDROME

Ocular Findings: Slight antimongoloid slant of the eyes.
Exotropia.

Associated Clinical Findings: Slightly beaked, straight nose.
Small skull.
Assymetry of the arch of the eyebrows.

Etiology: Familial.

Bibliography

Keitel, H.: Peculiar versus diagnostic facies in pediatrics. *Med. Sci.*, 17/6:47-58, 1966.
Kushnick, T.: Brachydactyly, facial abnormalities, and mental retardation. *Amer. J. Dis. Child.*, 111:96-98, 1966.

RUCKER'S SIGN

Ocular Findings: Sheathing of the retinal veins in the posterior pole.

Significance: Seen in about fifteen per cent of patients with multiple sclerosis.

Considered to be evidence of active perivasculitis in the retina.

Bibliography

Kearns, T. P.: Neuro-ophthalmology. *Annual Review. Arch. Ophthal.*, 76:729-755, 1966.
Archambeau, P. L.; Hollenhorst, R. W.; and Rucker, C. W.: Posterior uveitis as a manifestation of multiple sclerosis. *Mayo Clin. Proc.* 40:544-551, 1965.

RUD'S SYNDROME

Ocular Findings: Pigmentary retinopathy.

Associated Clinical Findings: Congenital ichthiosis.

Infantilism.

Epilepsy.

Polyneuritis.

Hyperchromic macrocytic anemia may be found.

Pathology: The brain cells show immaturity of the nerve cells as well as a decrease in actual number. A marked excess of oligodendroglia in the frontal cortex has been noted.

Etiology: May be related to the congenital neuroectodermal dysplasias as tuberous sclerosis and neurofibromatosis.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

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SABIN-FELDMAN SYNDROME

Ocular Findings: Chorioretinitis which may be unilateral or bilateral.

Associated Clinical Findings: Cerebral calcifications, but the methylene blue dye test in such cases is negative.

Pathology: The microscopic findings are similar to those seen in toxoplasmosis.

Etiology: Unknown.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

SABOURAUD'S SYNDROME

Ocular Findings: Keratosis pilaris (dryness, thickening and fragility of the lashes and brows).

Juvenile cataracts.

Associated Clinical Findings: Spindle-shaped thickened body hair, characterized by nodules on the hair shaft.

May have nail alterations.

May have teeth abnormalities.

Seen primarily in children.

Etiology: Familial.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

SAENGER'S SYNDROME

Synonyms: *Pupilotonia, Adie's Syndrome, Markus Syndrome.*

Ocular Findings: Pupil dilates and constricts very slowly to light and accommodation.

Reacts readily to miotics and mydriatics.

The dilated pupil is constricted with 2.5% mecholyl which has no effect on the normal pupil.

Associated Clinical Findings: Affects young women most frequently.

May have absent knee and ankle reflexes.

Etiology: Unknown.

Bibliography

Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.

SAINTON'S SIGN

Ocular Findings: The frontalis muscle contracts after the levator ceases to act in upward gaze.

Associated Clinical Findings: Graves' disease.

Increased basal metabolism.

Other systemic signs of hyperthyroidism.

SALUS' SIGN

Ocular Findings: Arterio-venous bridging with lateral deflection.

Significance: Seen in hypertension. One of the components of hypertensive retinopathy.

See Gunn's Sign.

SALZMANN'S DYSTROPHY

Synonym: *Nodular Dystrophy.*

Ocular Findings: Corneal degeneration following corneal disease, with degeneration and vascularization of the stroma and epithelium.

Dense whitish nodules are seen in the superficial stroma.

The epithelium may be raised over the nodules.
Always follows phlyctenular keratitis.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

SANFILIPPO SYNDROME

Synonym: *Mucopolysaccharidosis III*.

Similar Conditions: *Hurler's Syndrome, Hunter's Syndrome*.

Ocular Findings: Corneal clouding as "mild corneal opacities" may be seen.

Albinoid fundi have been described.

Associated Clinical Findings: Mental deterioration, severe by age five or six.

May have dwarfing, stiff joints, and hepatosplenomegaly, but these changes are minor as compared to Hurler's or Hunter's.

Etiology: Hereditary. Recessive.

Bibliography

Gills, J. P.; Hobson, R.; Hanley, W. B.; *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopoly-saccharidoses. *Arch. Ophthalm.*, 74:596-603, 1965.

Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. *Arch. Ophthalm.*, 74:516-520, 1965.

SANYAL'S CONJUNCTIVITIS

Ocular Findings: This is a peculiar type of actinomycetic infection due apparently to direct infection with dirt.

Seen primarily in India.

The disease is unilateral and characterized by a velvety chemotic appearance of the conjunctiva, without lymphatic involvement or systemic disturbance.

Clears up spontaneously in eight or ten weeks.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

SATTLE'S VEIL

Ocular Findings: Corneal edema.

Blurring of vision with the appearance of halos.

May progress to vesiculation.

Pathology: Epithelial edema due to oxygen lack. Usually produced by scleral contact lenses.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

SCHAUMANN'S SYNDROME

Synonyms: *Besnier-Boeck-Schaumann Syndrome, Boeck's Sarcoidosis.*

Ocular Findings: Granulomatous uveitis.

Band shaped keratitis.

Vitreous reaction.

Exudative retinopathy.

Associated Clinical Findings: Systemic sarcoidosis.

Lymphadenopathy.

Hilar nodes seen on x-ray.

Lack of energy and easy fatiguability.

Negative O.T. test.

Etiology: Unknown. Negroes primarily affected. Course is chronic with recurrences and spontaneous remissions.

Bibliography

Ozazewski, J. C. and Bennett, V.: Ocular sarcoidosis; report of 3 cases. *Amer. J. Ophthalm.*, 35:547-550, 1952.

SCHEIE SYNDROME

Synonym: *Mucopolysaccharidosis V.*

Similar Conditions: *Hurler's Syndrome, Hunter's Syndrome.*

Ocular Findings: Marked corneal clouding which may be present at birth, and is progressive.

Retinal pigmentary anomalies.

Associated Clinical Findings: Stiff joints.

Course facial features (but not approaching gargoylism).

Hirsutism.

Intelligence is normal or above normal.

Pathology: Urine is characterized by excessive amounts of chondroitin sulfate B.

Etiology: Congenital. Autosomal recessive.

Bibliography

Gills, J. P.; Hobson, R.; Hanley, W. B.; *et al.*: Electroretinography and fundus oculi findings in Hurler's disease and allied mucopolysaccharidoses. *Arch. Ophthalm.*, 74:596-603, 1965.

Goldberg, M. F.; Maumenee, A. E.; and McKusick, V. A.: Corneal dystrophies associated with abnormalities of mucopolysaccharide metabolism. *Arch. Ophthalm.*, 74:516-520, 1965.

SCHILDER'S DISEASE

Synonym: *Periaxialis Diffusa*.

Ocular Findings: Cortical blindness.

Eye grounds may be minimal. May have papilledema due to increased intracranial pressure.

May have nystagmus (nuclear or supranuclear in origin).

May have hemianopsia in occipital lobe involvement.

Associated Clinical Findings: Progressive mental deterioration.

Progressive spastic paralysis.

Irritability.

Tremors.

Deafness if temporal lobe is involved.

Affects young children.

Pathologic Etiology: Rare demyelinating disease. Lesions most frequent in the subcortical white matter. Male linkage.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Tamler, E. and Eissler, R.: The intraocular pathology in Schilder's disease. *Arch. Ophthalm.*, 65:514-516, 1961.

SCHIRMER'S SIGN

Findings: Failure of the lacrimal secretions in the resting (non-stimulated) eye to moisten a normal (usually fifteen millimeters) length of a filter paper test strip.

Significance: Indicates deficient lacrimation and is characteristic of Sjögren's syndrome or keratitis sicca.

Bibliography

Henderson, J. W.: Keratoconjunctivitis sicca; review with survey of 121 additional cases. *Amer. J. Ophthalm.*, 33:197-223, 1950.

SCHIRMER'S TEST

Findings: This is a test of lacrimal function used in the diagnosis of such conditions as neuroparalytic keratitis, filamentary keratitis, keratitis sicca, etc.

The test consists of placing the end of a strip of ordinary filter paper five by twenty-five millimeters in size, near the inner canthus in the lower conjunctival fornix.

Normally, about fifteen millimeters of the paper will be wet in five minutes (as measured from the fold at the lid margin).

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8, C. V. Mosby, St. Louis, Mo., 1965.

SCHNABEL'S ATROPHY

Ocular Findings: Cavernous atrophy of the optic nerve; a degenerative process seen in chronic glaucoma of long duration.

Accompanied by visual field defects of glaucoma.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

SCHNYDER'S CRYSTALLINE DYSTROPHY

Ocular Findings: Central or paracentral disciform opacity in the anterior layers of the cornea.

Usually bilateral.

Corneal sensitivity is only slightly reduced.

Visual acuity remains good.

Appears at birth or in early infancy.

Condition progresses extremely slowly.

Pathology: Opacity consists of crystals which are needle-shaped and whitish relucet to grayish in color. Possibly cholesterol.

Etiology: Hereditary. Dominant transmission.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

SCHÖNENBERG'S SYNDROME

Ocular Findings: Blepharophimosis.

Pseudoptosis.

Occasional epicanthus.

Associated Clinical Findings: Proportional dwarfism.

Congenital heart defect.

Etiology: Possibly familial.

Bibliography

Schönenberg, H.: Über ein neues Kombinationsbild multipler abartungen. (Minderwuchs, vitium cordis, beiderseitige congenitale ptose). *Ann. Pediat. (Basel)*, 182:229-240, 1954.

SÉZARY'S SYNDROME

Synonym: *Reticulosis Syndrome*.

Ocular Findings: Ectropion.

Thickening and edema of the lids.

Associated Clinical Findings: Generalized erythrodermia.

Intense pruritis.

Pigmentation and mottling of the skin.

Superficial lymphadenopathy.

Patchy alopecia.

Attacks of hyperhidrosis.

Occurs chiefly in elderly females.

Chronic course.

Pathology: Thought to be a disturbance of the reticuloendothelial system.

Bibliography

Alderson, W.; Barrow, G.; and Turner, R.: Sézary's syndrome. *Brit. Med. J.*, 1:256-260, 1955.

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

SFORZINI'S SYNDROME

Ocular Findings: Exophthalmos (non-thyroid).

Structural changes of the bony orbit.

Associated Clinical Findings: Stuttering.

Tall stature due to hyperplasia of the long bones.

Intestinal prolapses.

Normal metabolic rate.

Etiology: Felt to be on pituitary basis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

SHÄFER'S SYNDROME

Synonym: *Congenital Dyskeratosis*.

Similar Condition: *Jadassohn-Lewandowsky Syndrome*.

Ocular Findings: Corneal dystrophy.

Cataracts.

Associated Clinical Findings: Growth and mental retardation in childhood.

Keratosis palmo-plantaris.

Leukokeratosis of buccal mucosa.

Thickened nails.

Testicular hypoplasia.

Etiology: Congenital. Hereditary.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

SICHEL'S DISEASE

Synonyms: *Pseudoptosis, Sichel's Ptosis, Blepharochalasis.*

Ocular Findings: Fold or folds of skin hanging from the upper lid over the free lid margin.

Occurs in adults of all ages.

Once thought to be due to an accumulation of fat, but has been shown to be due to relaxation of facial bands which attach the levator to the skin of the lid.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

SIEGRIST SPOTS

Synonyms: *Siegrist's Sign, Siegrist Beads.*

Ocular Findings: Chains of pigmented spots, like a string of beads, along a sclerosed choroidal vessel.

Seen as a sequela of choroiditis.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

SIEGRIST'S SYNDROME**Ocular Findings:** Exophthalmos.

Progressive decrease in vision.

Granular pigmented spots in the choroid following the course of larger choroidal vessels.

Changes appear to be related to choroidal arteriosclerosis.

Associated Clinical Findings: Hypertension.

Albuminuria.

Etiology: Thought to be familial. Female sex-linkage.

Bibliography

Duke-Elder, Sir S.: *Textbook of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1941.

SIEMENS' DISEASE

Synonym: *Keratosis Follicularis*.

Ocular Findings: Follicular keratosis of the eyelids.

Piliary keratosis of the eyelashes with loss of lashes.

Thickening of the eyelids.

Blepharitis.

Ectropion.

Degenerative lesions of the cornea characterized by diffuse punctate epithelial changes and peripheral superficial vascularization in arches.

Photophobia and lacrimation.

Associated Clinical Findings: Piliary keratosis of the scalp, beard, and eyebrows.

Follicular keratosis of the nape of the neck, the ears, the vertebral groove, palms of the hands, and the soles of the feet.

Etiology: Autosomal dominant inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

SJÖGREN'S SYNDROME

Ocular Findings: Congenital zonular or polar cataract.

Associated Clinical Findings: Oligophrenia.

Hereditary ataxia.

Other congenital anomalies such as small stature, scoliosis, genu valgum and defects of the fingers and toes.

Etiology: Hereditary. Autosomal recessive.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

SJÖGREN'S SYNDROME

Synonyms: *Gougerot-Sjögren Syndrome, Keratoconjunctivitis Sicca.*

Ocular Findings: Blepharoconjunctivitis.

Deficient lacrimal secretion with a positive Schirmer's sign.

Vision may be blurred.

Photophobia.

Stringy mucoid discharge.

Filimentary keratoconjunctivitis.

Associated Clinical Findings: Dryness of the mouth.

Polyarthrititis.

Scleroderma-like skin changes.

Muscle weakness.

Areflexia.

Alopecia.

Pathology: Deficient lysozyme production. Atrophic changes in the lacrimal glands.

Etiology: Unknown.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

Henderson, J. W.: Keratoconjunctivitis sicca; review with survey of 121 additional cases. *Amer. J. Ophthalm.*, 33:197-223, 1950.

SJÖGREN-LARSSON SYNDROME

Synonyms: *Xerodermal Idiocy, Torsten-Sjögren Syndrome.*

Ocular Findings: Macular degeneration, usually bilateral.

Associated Clinical Findings: Congenital ichthiosis.

Oligophrenia.

Spastic paralysis of the lower extremities.

Etiology: Familial recessive. Consanguinity may be a factor.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

SLUDER'S SYNDROME

Synonym: *Sphenopalatine Ganglion Syndrome.*

Ocular Findings: Ocular hyperemia.

Lacrimation.

Orbital and periorbital neuralgia.

Associated Clinical Findings: Unilateral nasopharyngeal congestion with rhinorrhea.

Radiation of neuralgia to the nose, palate, temple, ears and neck.

Etiology: Irritative lesion of the sphenopalatine ganglion.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

SOEMMERING'S RING

Ocular Findings: Lens remnants enclosed in portions of the lens capsule.

Arranged in the periphery in the area of the lens equator to form a ring with an open or partially open pupillary area.

The pupillary area may contain remnants of the anterior or posterior capsule.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

SORSBY'S DISEASE

Synonym: *Sorsby's Inflammatory Hereditary Degeneration of the Macula.*

Ocular Findings: Macular degeneration of hemorrhagic-exudative type.

Onset between age 20 to 40 years.

Marked by the appearance in the macular region of edema, hemorrhage, and exudates.

These lesions, which are reminiscent of a process of inflammation or arteriosclerosis, develop gradually into chorioretinal atrophy which extends toward the periphery and may show pigmentary degeneration.

Etiology: Familial. Dominant transmission.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

SORSBY'S SYNDROME

Similar Conditions: *Biemond's Syndrome, Bardet-Biedl's Syndrome.*

Ocular Findings: Coloboma of the macula.

Atypical pigmentary dystrophy.

Associated Clinical Findings: Bradydactyly.

Syndactyly or polydactyly.

Etiology: Familial. Probable recessive transmission.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

SPANNLANG-TAPPEINER SYNDROME

Ocular Findings: Zonular opacities of the cornea.
Cataracts have been reported.

Associated Clinical Findings: Keratosis palmo-plantaris.
Hyperhidrosis.
Partial alopecia.

Etiology: Hereditary. Recessive.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

SPIELMEYER-VOGT SYNDROME

Synonyms: *Batten-Mayou Disease, Juvenile Amaurotic Family Idiocy.*

Ocular Findings: Diminished vision.
Onset between age five and eight.
Ocular findings resemble retinitis pigmentosa.
Progresses to primary optic atrophy and complete blindness.

Associated Clinical Findings: Pathologic behavior changes.
Ataxia.
Tremors.
Upper and lower motor neuron palsies.
Complete paralysis and dementia occur in the terminal stages.

Etiology: Heredofamilial. Simple recessive.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

SPURWAY'S SYNDROME

Synonym: *Brittle Bones—Blue Sclera Syndrome.*

Similar Conditions: *Lobstein's Syndrome, Van der Hoeve's Syndrome.*

Ocular Findings: Blue sclera, of a relatively uniform color because of the thinness and transparency of the tissue.

May have associated megalocornea.

Associated Clinical Findings: Bony fragility resulting in multiple and repeated spontaneous fractures.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

STARGARDT'S DISEASE

Synonym: *Juvenile Hereditary Disciform Macular Degeneration.*

Similar Condition: *See Khunt-Junius Syndrome.*

Ocular Findings: Disciform macular degeneration.

Usual onset age, eight to fifteen; more often at age of puberty.

Associated with peripheral pigmentary degenerative retinal lesions without hemeralopia.

Begins with the disappearance of the foveal reflex or slight macular edema; then small spots appear, grayish to orange in color, and between these spots a network of coalescing pigment deposits form and spread over the entire macular area.

The macular area becomes more strongly pigmented with time and finally becomes atrophic and transparent enough to allow some choroidal vessels to be seen.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis Mo., 1961.

STEINERT'S DISEASE

Synonym: *Steinert's Myotonic Dystrophy.*

Ocular Findings: Posterior subcapsular stellate cataract.

Atypical tapetoretinal degeneration.

Trophic disorders of the cornea.

Convergence deficiencies with diplopia.
Ptosis.

Associated Clinical Findings: Onset in the third or fourth decade of life.
Myopathies of the face involving the masseters and sternomastoid muscles.
Muscular atrophy of the arm, hand, and thigh muscles.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

STELLWAG'S SIGN

Ocular Findings: Retraction of the upper lid associated with infrequent and incomplete blinking.
This sign is present in less than half of the cases of hyperthyroidism.

Associated Clinical Findings: Graves' disease.
Increased basal metabolism.
Tachycardia.
Nervousness.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

STEVENS-JOHNSON SYNDROME

Synonyms: *Dermatostomatitis, Erythema Multiforme.*

Similar Condition: *Hebra's Disease.*

Ocular Findings: Purulent pseudomembranous conjunctivitis.
Symblepharon.
Hypopyon.
Bulbar and conjunctival chemosis.

Keratitis and iritis.

May progress to partial or total blindness.

Associated Clinical Findings: Stomatitis.

Severe generalized maculopapular rash (erythema multiforme).

Rhinitis.

Acute respiratory infections.

Urethritis and balanitis.

General malaise.

May be confused with Reiter's Syndrome.

Etiology: Unknown. May be partly due to drug allergy.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Robinson, H. M., Jr. and McCrumb, F. R., Jr.: Comparative analysis of mucocutaneous—ocular syndromes; report of 11 cases and review of literature. *Arch. Dermat. and Syph.*, 61:539-560, 1950.

STILL'S DISEASE

Ocular Findings: Slow, chronic iridocyclitis.

Band-shaped corneal opacity (calcareous incrustation of Bowman's membrane).

May be complicated by cataract.

Associated Clinical Findings: Juvenile polyarthritis, with little joint damage.

Splenomegaly.

Generalized enlargement of the lymph nodes.

Occurs typically in female children.

Prognosis poor.

Many children with Still's disease die before the eye lesions can develop.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Smiley, W.: Iridocyclitis in Still's disease. *Trans. Ophthal. Soc. UK*, 85:351-356, 1965.

STILLING-DUANE-TURK'S SYNDROME

Synonym: *Duane's Syndrome.*

Ocular Findings: Limitation or absence of abduction.

Retraction of the eyeball with narrowing of the palpebral fissure on adduction of the eye.

May have vertical deviation due to enhanced action of the inferior oblique muscle.

May have heterochromia iridis.

Bilateral in fifteen to twenty per cent of cases.

Occurs more in females.

Pathology: Due to congenital deformity of the eye muscles and their tendons of insertion.

Etiology: Thought to be familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

STOCK-SPIELMEYER DISEASE

Synonyms: *Vogt-Spielmeyer Disease, Batten's Disease, Juvenile Amaurotic Family Idiocy.*

Ocular Findings: Progressive amaurosis.

Cherry red macula.

Retinal degeneration resembling retinitis pigmentosa.

Progressive primary optic atrophy.

Associated Clinical Findings: Onset at five to eight years of age.

Pathologic personality changes.

Mental regression.

Mortality usually by age eighteen.

Pathology: Cerebroretinal degeneration with degeneration of the rods and cones.

Etiology: Familial. Recessive.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

STOCKER'S LINE

Ocular Findings: Rarely observed brownish pigmented line running parallel with and just anterior to the head of a pterygium in the superficial cornea.

Clinically resembles the Hudson-Stahli Line.

Pathology: Felt to be due to iron deposits within the corneal epithelium and may represent a deviation of the line of normal iron deposition in response to pathological alteration of the superficial cornea.

Bibliography

Gass, J. D.: The iron lines of the superficial cornea; Hudson-Stakli line, Stocker's line and Fleischer's ring. *Arch. Ophthal.*, 71:348-358, 1964.

STREIFF'S SYNDROME

Synonym: *Posterior Marginal Dysplasia of the Cornea.*

Similar Condition: *Axenfeld's Syndrome.*

Ocular Findings: Whitish glassy stria, annular in configuration and localized in Descemet's membrane.

Separated from the corneal limbus by a relatively clear zone.

Associated with defects of the iridocorneal angle.

May have hyaline membrane bridging from the peripheral iris and adherent to the posterior surface of the cornea.

May have heterochromia.

May have corectopia.

May have ectropion uvei.

Pathology: Hypertrophy of Schwalbe's ring. Predisposes to glaucoma because of the presence of goniosynechiae.

Bibliography

Streiff, E. B.: Dysplasie marginale postérieure de la cornée (embryontoxon posterius

Axenfeld) dans le cadre des malformations irido-cornéennes. *Ophthalmologica* (Basel), 118:815-827, 1949.

STRÜMPPELL-LORRAIN DISEASE

Synonym: *Familial Spasmodic Paraplegia.*

Ocular Findings: Strabismus.

Pupillary defects.

Juvenile macular degeneration of Staargardt type.

May have optic atrophy.

Associated Clinical Findings: Generally appears in the second decade.

Associated with spasmodic hypotonic paresis of pyramidal origin,
without muscular atrophy, ataxia, or sensory disturbances.

Etiology: Familial. Regular dominant or recessive.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

STURGE-WEBER SYNDROME

Synonyms: *Sturge-Weber-Kalischer-Dimitri Syndrome, Encephalotrigeminal Angiomatosis.*

Ocular Findings: Angioma of the choroid.

May have hydrophthalmos.

May have glaucoma if port-wine hemangioma involves the eyelids.

Occasional port-wine hemangioma of the lids.

Occasional ablatio retinae.

Associated Clinical Findings: Usually unilateral nevus flammeus (port-wine hemangioma of the skin), usually in the area of the fifth nerve distribution.

Meningeal angiomas.

Convulsions may occur.

Mental retardation may be found.

Pathology: Intracranial calcification on x-ray.

Etiology: Felt to be a partial trisomy of a D chromosome. Simple recessive transmission.

Bibliography

- Berkow, J. W.: Retinitis pigmentosa associated with Sturge-Weber syndrome. *Arch. Ophthalm.*, 74:72-76, 1966.
Francois, J.: Chromosome abnormalities and ophthalmology. *J. Pediat. Ophthalm.*, 1:5-18, 1964.
Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

SUKER'S SIGN

Ocular Findings: Inability to maintain fixation on extreme lateral rotation.

An inconstant and infrequently noted sign of hyperthyroidism.

Associated Clinical Findings: Graves' disease.

Increased basal metabolism.

Trembling.

Other signs of hyperthyroidism.

Bibliography

- Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

SWAN'S BLIND SPOT SYNDROME

Ocular Findings: In esotropias of ten to fifteen degrees (twenty to thirty prism diopters), the use of the physiologic blind spot in the deviating eye as the suppression area.

This may actually be a compensatory convergence by means of which an individual with strabismus is able to avoid diplopia.

As the individual converges to help his accommodation, the physiologic blind spot of the squinting eye receives the "second image" of the object fixed and thus prevents diplopia.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
Swan, K. C.: Squint syndrome. *Arch. Ophthalm.*, 37:149-154, 1947.

SWIFT'S DISEASE

Synonyms: *Feer's Syndrome, Infantile Acrodynia.*

Ocular Findings: Photophobia with or without conjunctivitis.
Proptosis in about fifty per cent of cases.

Associated Clinical Findings: In older infants, erythema, eruptions, and hyperkeratosis of the skin.

Weakness.

Irritability.

Profuse sweating.

Hypertension.

Cyanosis of the extremities.

Etiology: Uncertain.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

SYMONDS' SYNDROME

Synonyms: *Otitic Hydrocephalus Syndrome, Serous Meningitis Syndrome.*

Ocular Findings: Diplopia due to sixth nerve palsy.

Papilledema.

Optic atrophy.

Retinal hemorrhages.

Associated Clinical Findings: High elevation of spinal fluid pressure.

Otitis media.

Mastoiditis.

Meningitis.

Brain abscess may occur.

Headaches.

Drowsiness.

Occurs in childhood.

Prognosis good for spontaneous recovery.

Bibliography

Symonds, C. P.: Otitic hydrocephalus; report of 3 cases. *Brit. Med. J.*, 1:53-54, 1932.

T

TAKAYASU-OHNISHI DISEASE

Synonyms: *Takayasu's Disease, Pulseless Disease.*

Ocular Findings: Orthostatic or fluctuating blurred vision and photopsia.

May be monocular or binocular.

Tortuosity and distention of the retinal veins with increased visibility of capillary anastomoses and capillary microaneurysms.

May have neovascularization and atrophy of the iris.

May have cataracts.

May have pigmentary abnormalities of the retina.

Associated Clinical Findings: Syncopal attacks.

Convulsions.

Loss of radial artery pulse.

May have fever.

Pathologic Etiology: Signs and symptoms due to carotid and aortic constriction.

Giant-cell arteritis with periarteritis of the aortic arch.

Bibliography

Hedges, T. R.: The aortic arch syndromes. *Arch. Ophthalm.*, 71:28-34, 1964.

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

TAY-SACHS' DISEASE

Synonym: *Infantile Amaurotic Family Idiocy.*

Ocular Findings: Bilateral cherry red spot in the macula.

Blindness due to optic atrophy with absent pupillary light reflex.

May have nystagmus.

May have strabismus.

May have fine diffuse punctate corneal opacities.

Associated Clinical Findings: Onset three to six months after birth.

Progressive dementia.

Idiocy.

Somnolence.

Epilepsy.

Muscular pareses.

Mainly in Jewish children (reports vary from sixty to eighty five per cent).

Pathology: Lipoid degeneration of ganglion cells of the retina and central nervous system.

Etiology: Simple recessive inheritance.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Dyken, P. and Zernan, W.: A clinical, pathological, and genetic study of an unusual form of Tay-Sachs' disease with macular degeneration in the family. *Survey Ophthalm.*, 11:238-240, 1966.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

TERRIEN'S DISEASE

Synonyms: *Terrien's Marginal Degeneration of the Cornea, Peripheral Furrow Keratitis.*

Ocular Findings: A rare bilateral condition starting with opacification and vascularization of the cornea in the periphery.

Progresses by degeneration of the stroma to the formation of a gutter or furrow.

Finally results in ectasia of the peripheral cornea.

May involve part or all of the circumference of the cornea.

Most (about three-fourths) occur in males.

Though most occur in older adults, it can occur at any age.

May resemble arcus senilis in early stages but then becomes vascularized.

Etiology: Unknown.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

TERRY'S SYNDROME

Synonym: *Retrolental Fibroplasia*.

Ocular Findings: Leukocoria due to grayish opacities in the anterior vitreous.

Myopia.

Nystagmus.

Strabismus.

Retinopathy.

May be complicated by glaucoma.

Course is frequently progressive.

May be monocular or binocular.

Pathology: Mass of vascular embryonic retinal connective tissue and vessels behind the lens.

Etiology: Due to high oxygen concentration used with premature infants immediately after birth.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Terry, T. L.: Extreme prematurity and fibroblastic overgrowth of persistent vascular sheath behind each crystallin lens; preliminary report. *Amer. J. Ophthal.*, 25:203-204, 1942.

TERSON'S DISEASE

Ocular Findings: Ocular venous stenosis, and venous saccular dilations. Cloudy vitreous.

Associated Clinical Findings: Multiple cerebral aneurysms.

Subarachnoid hemorrhages, venous, due to aneurysm rupture.

Males mainly affected.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

THOMPSON'S DISEASE

Synonym: *Thompson's Congenital Optic Atrophy.*

Similar Condition: *Leber's Disease.*

Ocular Findings: Congenital amaurosis due to optic atrophy.

Pupillary reactions slight or absent.

Pendular nystagmus.

May have associated cataracts.

Pathology: Dysgenesis of the neuroepithelium.

Etiology: Autosomal recessive transmission.

Bibliography

Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

Thompson, A. H. and Cashell, G. T. W.: Pedigree of congenital optic atrophy embracing 16 affected cases in 6 generations. *Proc. Roy. Soc. Med.*, 28:1415-1425, 1935.

THOMSEN'S DISEASE

Synonym: *Congenital Muscular Dystrophy.*

Similar Condition: *Steinert's Disease.*

Ocular Findings: May have ptosis.

Difficulty in quickly turning the eyes in any direction and may have difficulty opening the eyes.

Cataracts are never found in Thomsen's disease.

Associated Clinical Findings: Characterized generally by a systemic hypertrophic myotonia with slow relaxation of muscular contraction.

Etiology: Familial. Dominant inheritance.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

THOMSON'S SYNDROME

Synonym: *Congenital Poikiloderma.*

Similar Condition: *Rothmund's Syndrome. May be identical.*

Ocular Findings: Bilateral congenital cataracts.

High myopia.

Sensitivity to sunlight.

Associated Clinical Findings: Erythematous swelling of the face, buttocks and extremities, followed by atrophy of the skin and pigmentary changes.

Alopecia and loss of the eyebrows.

Etiology: Unknown.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Thomson, M. S.: Poikiloderma congenitale. *Brit. J. Derm.*, 48:221-234, 1936.

THYGESON'S DISEASE

Ocular Findings: Superficial punctate epithelial keratitis.

No associated conjunctivitis.

Coarse irregular surface with minimal stromal involvement.

Runs a chronic or recurrent course of many years.

Non-infectious.

Etiology: Unknown.

Bibliography

Jones, B. R.: Differential diagnosis of punctate keratitis. *Int. Ophthalm., Clin.*, 2:591-611, 1962.

Thygeson, P.: Further observations on superficial punctate keratitis. *Arch. Ophthalm.*, 66:158-162, 1961.

TILLING-WERNICKE SYNDROME

Ocular Findings: Pseudonuclear ophthalmoplegia.

Paresis of volitional eye movements.

Associated Clinical Findings: Dissociated pareses of the facial muscles, the muscles of swallowing, etc.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

TOGBY'S DISEASE

Synonym: *Vernal Catarrh.*

Ocular Findings: Recurrent seasonal superficial keratitis.
Stringy exudates.
Accompanied by itching.

Etiology: Allergic.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

TOURNAINE'S DISEASE

Synonym: *Systemic Elastorrhexia.*

Similar Condition: *Groenblad-Strandberg Syndrome.*

Ocular Findings: Angeoid streaks of the retina in about seventy per cent of cases.

Associated Clinical Findings: Cutaneous pseudoxanthoma elasticum.
Usually begins in the third decade.
Cardiovascular lesions such as premature arteriosclerosis and hypertension, peripheral arteritis and myocarditis.

Pathology: Elastic degeneration of the middle coats of the arteries.

Etiology: Hereditary.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

TOURNAY'S REACTION

Synonym: "*Normal Anisocoria.*"

Ocular Findings: Inequality of the pupils when the eyes are turned strongly to either side, with dilation of the *abducting* eye.

The *adducting* eye may also constrict.

This is opposite to the pathologic abduction phenomenon of Behr.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

TRANTAS' SPOTS

Synonym: *Horner-Trantas Spots.*

Ocular Findings: Chalky concretions appearing as small white specks in the tarsoconjunctiva.

Seen in the later stages of vernal catarrh.

They are the end products of papillary hypertrophy.

Pathology: Concretions are made up of degenerated eosinophiles and calcium.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

TREACHER-COLLINS' SYNDROME

Synonym: *Incomplete Mandibulo-facial Syndrome.*

Similar Condition: *Francheschetti's Syndrome.*

Ocular Findings: Colobomas of the outer one-third of the lower lids.

Absence of the lower puncta.

Partial alopecia ciliaris medial to the coloboma.

Oblique palpebral fissure with lower outer canthus.

Associated Clinical Findings: Micrognathia.

Auricular appendages.
Occasionally absence of the external ear.
Hypoplastic zygomatic arch.

Etiology: Unknown. Possible arrested fetal development.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
Harrison, S. H.: Treacher-Collins' syndrome. *Brit. J. Plast. Surg.*, 3:282-290, 1951.
Keitel, H.: Peculiar versus diagnostic facies in pediatrics. *Med. Sci.*, 17/6:47-58, 1966.

TURK'S LINE

Ocular Findings: Vertical line of pigment granules which collect on the corneal endothelium in the center of the lower one-third of the cornea.

Arrangement in a vertical line is due to the normal convection currents of the aqueous.

The pigment granules are shed from the iris.

In mild forms, and in old people, these are often normal senile changes. They are more marked, more frequent, and occur at an earlier age, however, in pathologic states, especially myopia, diabetes, glaucoma, and after intraocular surgery.

Bibliography

- Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

TÜRK'S SYNDROME

Synonym: *Türk's Disease. May be spelled Tuerk.*

Similar Condition: *Duane's Syndrome.*

Ocular Findings: Fibrous degeneration of the external rectus muscle with limitation of abduction of the affected eye beyond the mid-line and retraction of the globe upon adduction.

This limitation of muscular function is one of the components of

Duane's Retraction Syndrome; hence, Türk's Syndrome is regarded as an incomplete form of the latter.

Etiology: Birth injury has been suggested as one of the more likely causes.

Bibliography

- Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.
Kestenbaum, A.: *Clinical Methods of Neuro-ophthalmologic Examination*, 2nd ed. Grune and Stratton, New York, N.Y., 1961.

TURNER'S SYNDROME

Similar Condition: *Bonnevie-Ullrich Syndrome*.

Ocular Findings: Strabismus.

Ptosis.

May have cataracts.

May have corneal nebulae.

Eight per cent are color blind (in the normal population, females are color blind in only 0.4%), the same ratio as the normal male.

Epicanthus.

Associated Clinical Findings: Retarded growth.

Retarded sexual development.

May have webbing of the neck.

Absence of chest hair.

Infantilism.

Rudimentary ovaries.

Pathologic Etiology: Patients are phenotypically female but the sex chromatin is male.

Affected females have 45 chromosomes; 44 autosomes and one X chromosome.

Bibliography

- Francois, J.: Chromosome abnormalities and ophthalmology. *J. Pediat. Ophthalm.*, 1:5-18, 1964.
Lessel, S. and Forbes, A. P.: Eye signs in Turner's syndrome. *Arch. Ophthalm.*, 76:211-213, 1966.

U

UHTHOFF'S SIGN

Synonym: *Uhthoff's Symptom.*

Ocular Findings: Impaired visual acuity is further reduced by heat or exercise in cases of multiple sclerosis.

Associated Clinical Findings: Multiple sclerosis.

Significance: Since exercise and physical exertion have been noted to influence the visual symptoms of multiple sclerosis, an exercise test may be of some help in cases of optic neuropathy when the etiology remains in doubt.

Bibliography

- Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.
Goldstein, J. E. and Cogan, D. G.: Exercise and the optic neuropathy of multiple sclerosis. *Arch. Ophthalm.*, 72:168-170, 1964.

ULLRICH'S SYNDROME

Synonyms: *Dyscranio-pygo-phalangea*, *Ullrich-Feichtiger Syndrome*.

Ocular Findings: Bilateral anophthalmos or microphthalmos.
Blepharophimosis.
Chorioretinal colobomas.
Aniridia.

Associated Clinical Findings: Cranial deformities characterized by a bradycephalic or acrocephalic skull with a broad nose and small mandible.

Skeletal deformities such as spina bifida and polydactyly.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

ULLRICH-FREMERIEY-DOHNA SYNDROME

Synonyms: *Dyscephalia Oculo-mandibulo-facialis*, "Bird Face Syndrome."

Ocular Findings: Blue sclera.

Congenital cataracts.

Occasional microphthalmos.

Associated Clinical Findings: "Triangular-shaped" microcephalus.

Microstomia.

Teeth anomalies.

Retarded growth.

Hypotrichosis.

Skin atrophy.

Spinal cord anomalies.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

ULLRICH-TURNER SYNDROME

Synonym: *Pterygonuclear Infantilism*.

Ocular Findings: Ptosis.

Congenital cataracts.

Retinal pigmentary degeneration.

Associated Clinical Findings: "Sphinx face."

Developmental anomalies.

Pterygium colli.

Gonadal dysgenesis.

Infantilism.

Etiology: Congenital.

Bibliography

von Grósz, C.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

URBACH-WIETHE DISEASE

Synonym: *Lipoid Proteinosis*.

Ocular Findings: At the free margins of the eyelids, multiple, small, juxtaposed nodules occur, resembling milia with an appearance like fatty "blisters."

Moniliform blepharitis may be seen.

May result in loss of the eyelashes.

Associated Clinical Findings: Nodular and hyperkeratotic eruptions scattered over the head and extremities.

Hoarseness due to similar nodular changes on the vocal cords.

Pathology: Nodules are subepithelial lipoid deposits.

Etiology: Hereditary.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

USHER'S SYNDROME

Ocular Findings: Pigmentary Degeneration of the Retina (Retinitis Pigmentosa).

Associated Clinical Findings: Congenital or early developmental deafness.

May be associated with aphasia.

Etiology: Simple recessive inheritance.

Bibliography

Berkow, J. W.: Retinitis pigmentosa associated with Sturge-Weber syndrome. *Arch. Ophthalmol.*, 75:72-76, 1966.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

UYEMURA'S SYNDROME

Ocular Findings: Hemeralopia.

Xerosis oculi.

Yellow-white dots on the retina with the appearance of fundus albi-punctatus.

Associated Clinical Findings: Xerodermia.

Etiology: Avitaminosis A.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

V**VAN BOGAERT-DIVRY SYNDROME**

Synonym: *Diffuse Cortico-meningeal Angiomatosis. Compare with Sturge-Weber and Rothmund's Syndromes.*

Ocular Findings: Hemianopsias.

Retinal hemangiomas may be seen.

Associated Clinical Findings: Severe mobility defects.

Dementia.

Epilepsy.

Poikiloderma.

Pathology: Cortico-meningeal angiomas.

Etiology: Recessive sex-linked.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

VAN BOGAERT-SCHERER-EPSTEIN SYNDROME

Synonym: *Familial Hypercholesterolemia Syndrome.*

Ocular Findings: Xanthelasma.

Juvenile cataracts.

Associated Clinical Findings: Multiple xanthomatosis.

Defects of the central nervous system.

Hypercholesterolemia.

Etiology: Familial.**Bibliography**

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

VAN DER HOEVE'S SYNDROME

Synonyms: *Spurway's Syndrome, Brittle Bones Disease, Osteogenesis Imperfecta.*

Ocular Findings: Blue sclera (due to thinning of the sclera).

Cornea may be thinned leading to megalocornea of keratoconus.

May have telecanthus.

Associated Clinical Findings: Brittle bones resulting in multiple fractures and dislocations.

Hyperflexibility of ligaments.

Dental defects.

Deafness in about sixty per cent of cases, bilateral and progressive; usually appearing after age twenty.

Skin may appear atrophic due to the failure to develop collagen.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Ruedemann, A.: Osteogenesis imperfecta congenita and blue sclerotics; clinico-pathologic study. A.M.A. Arch. Ophthal., 49:6-16, 1953.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

VAN TRIGT'S SIGN

Ocular Findings: Pulsation of the retinal venules on the optic disc.

Significance: Evidence of a low intraocular pressure. A normal finding.

Bibliography

Duke-Elder, Sir S.: *Textbook of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1941.

VASQUEZ'S DISEASE

Synonyms: *Polycythemia Vera*, *Cyanosis Retinae*.

Ocular Findings: Cyanotic fundus.

Dark, dilated, tortuous veins.

Dark arteries.

Hyperemic, edematous optic disc.

Cyanotic conjunctiva.

Associated Clinical Findings: Polycythemia, usually beginning between thirty and fifty years of age.

Splenomegaly.

Digestive dysfunctions.

Ocular findings may also be seen with congenital heart disease and Ayerza's disease (pulmonary arteriosclerosis).

Pathology: Progressive polyglobism with a tendency to hemorrhages and to venous thrombosis.

Etiology: Hereditary tendency.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

VILDERVANCK'S SYNDROME

Synonym: *Oculocervical Craniostenosis Syndrome*.

Ocular Findings: Congenital bilateral abducens paresis.

Bulbar retraction.

Heterochromia iridis.

Associated Clinical Findings: Patient is deaf and dumb.

Teeth anomalies.

Congenital short, webbed neck with immobility and torticollis similar to Klippel-Feil Syndrome.
Mental deficiency.

Etiology: Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

VILLARET'S SYNDROME

Ocular Findings: Horner's syndrome; ptosis, miosis, and relative enophthalmos.

Associated Clinical Findings: Unilateral paralysis of the ninth, tenth, eleventh, twelfth cranial nerves, the cervical sympathetic chain and occasionally fibers of the seventh cranial nerve.

Paralysis of the soft palate, pharynx, and vocal cords.

Loss of taste of the posterior one-third of the tongue.

Paralysis of the sternocleidomastoid and trapezius muscles.

Pathology: Lesion in the retroparotid space. May be traumatic or neoplastic.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Pollock, L.: Multiple cranial nerve palsy due to extracranial disease. JAMA, 78:502-503, 1922.

VOGT'S CATARACT

Synonym: *Frosted Cataract*.

Ocular Findings: In the superficial layers of the embryonic nucleus there are small whitish irregular scintillating opacities interspersed with fine crystalline needles haphazardly arranged.

Vision is reduced.

Etiology: Probable dominant inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

VOGT'S FLOURY CORNEA

Synonym: *Cornea Farinata*.

Ocular Findings: Corneal changes characterized by small pointlike opacities in the deep layers of the parenchyma, constituting a senile or presenile alteration of the cornea.

Develops in the fourth to the sixth decades.

Etiology: Familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

Pippow, G.: Zur erbbedingtheit der cornea farinata (mehlstaubartige hornhautdegeneration). Graefe Arch. Ophthal., 44:276-279, 1941.

VOGT'S MOSIAC DEGENERATION

Ocular Findings: Primarily affects Bowman's membrane.

Has the granular appearance of lizard skin or crocodile skin.

Round or polygonal grayish elements are separated by darker intervals, forming a kind of mosaic.

Occupies the central third of the area of the membrane.

May appear at any age.

Etiology: Familial. Autosomal dominant.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

VOGT'S SIGN

Synonym: *Vogt's Glaukoma Flecken*.

Ocular Findings: Irregular whitish flects on the anterior lens surface.

Seen in prolonged subacute glaucoma.

Bibliography

Shaffer, R. N.: *Stereoscopic Manual of Gonioscopy*. C. V. Mosby, St. Louis, Mo., 1962.

VOGT-KOYANAGI SYNDROME

Similar Condition: *Harada's Syndrome*.

Ocular Findings: Severe, bilateral anterior uveitis.

Reduced vision.

May have secondary glaucoma.

Associated Clinical Findings: Alopecia.

Poliosis.

Vitiligo.

Dysacusia or tinnitus.

Mostly occurring in dark races like the Japanese.

Bibliography

Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

VOGT-SPIELMEYER DISEASE

Synonyms: *Juvenile Amaurotic Family Idiocy*, *Stock-Spielmeyer Disease*, *Batten-Mayou Disease*.

Similar Conditions: *Tay-Sachs' Disease*.

Ocular Findings: Cherry red spot in the maculas.

Blindness with absent pupillary light reflex.

Vessels and optic disc appear normal early; later the disc becomes pale due to optic atrophy.

Usual onset from age five to eight.

Fundus may resemble retinitis pigmentosa.

Associated Clinical Findings: Progressive dementia.

Epileptiform attacks.

Extrapyramidal symptoms.

Rigid and bent posture.

Ataxic gait.

Etiology: Autosomal recessive inheritance.

Bibliography

- Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.
Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Hoffman, J.: Pigmentary retinal lipid neuronal heredodegeneration (Spielmeyer-Vogt disease); neuro-ophthalmologic considerations. *Amer. J. Ophthal.*, 42:15-21, 1956.

VON GIERKE'S DISEASE

Synonym: *Glycogen Storage Disease*.

Ocular Findings: Cloudiness of the marginal areas of the cornea.
Findings are bilateral.

Associated Clinical Findings: This is a congenital disease due to the accumulation of glycogen, which by an inborn error of metabolism cannot be available as a source of glucose.

It results in hypoglycemia and the accumulation of glycogen in certain body tissues such as the liver, heart, and muscles.

Etiology: Hereditary. Dominant.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.
Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

VON GRAEFE'S DISEASE

Synonym: *Chronic Progressive Ophthalmoplegia*.

Findings: Slowly progressive bilateral paralysis of all the extraocular muscles.

Internal ophthalmoplegia occurs only in the end stages.

Associated Clinical Findings: Systemic changes characteristic of the underlying disease.

Pathologic Etiology: May be seen in syphilis, multiple sclerosis, progressive muscular dystrophy, and progressive spinal muscular atrophy.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

VON GRAEFE'S SIGN

Synonym: *Lid Lag*.

Ocular Findings: The upper lid pauses, then follows the eye movement downward when the patient follows an object being moved downward.

Associated Clinical Findings: Graves' disease.

This is a relatively common sign of the congestive type of thyroid disease.

Other systemic signs of thyroid dysfunction.

Clinical Note: Though lid lag and lid retraction are not the same thing, many authors use the terms interchangeably and thus indentify both under the eponym Graefe or Von Graefe. Lid retraction is more properly termed Dalrymple's sign.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Haessler, F. H.: *Eye Signs in General Diseases*. Charles C Thomas, Springfield, Ill., 1960.

VON HERRENSCHWAND'S SYNDROME

Synonym: *Sympathetic Heterochromia*.

Similar Condition: *Horner's Syndrome*.

Ocular Findings: Unilateral heterochromia iridis.

Miosis.

Ptosis.

Enophthalmos (Horner's syndrome).

Associated Clinical Findings: Decreased sweating on ipsilateral side of the face.

Pathologic Etiology: Sympathetic palsy from cervical rib, thyroid tumor, apical tubercular nodules, etc.

Bibliography

- Geeraets, W.: *Ocular Syndromes*. Lea and Febiger, Philadelphia, Pa., 1965.
Mann, I.: *Developmental Abnormalities of the Eye*. Cambridge University Press, London, England, 1937.

VON HIPPEL'S ULCER

Synonym: *Posterior Ulcer of Von Hippel*.

Ocular Findings: Congenital absence of Descemet's membrane.
Corneal clouding may be seen.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

VON HIPPEL-LINDAU SYNDROME

Synonyms: *Lindau-Von Hippel Syndrome, Retinocerebral Angiomatosis*.

Similar Condition: *Von Hippel's Disease (see below)*.

Ocular Findings: Hemangiomas of the retina.

May have hemorrhages and exudates, and in severe cases, retinal detachment.

May have iris angiomatosis.

May be complicated by secondary glaucoma.

When the findings are confined to the retina, the process is called Von Hippel's disease.

Associated Clinical Findings: Cerebellar angiomas (usually cystic).

Polycystic pancreas and kidneys.

Ataxia.

Epilepsy.

More common in young men.

Etiology: Familial in about twenty per cent of cases.

Bibliography

- Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Joe, S. and Spencer, W. H.: Von Hippel-Lindau disease. *Arch. Ophthal.*, 71:508-509, 1964.

Segal, N.; Fori-Maté, I.; and Bologa, E.: Difficulties in the diagnosis of retinal angiomas (von Hippel's disease) in the early stage. *Oftalmologica*, 9:45-90, 1965.

VON RECKLINGHAUSEN'S DISEASE

Synonym: *Neurofibromatosis*.

Ocular Findings: Glaucoma which may be congenital.

Ptosis.

Exophthalmos may be seen.

Gliomas of the optic nerve.

Papilledema.

Retrobulbar neuritis and atrophy of the optic disc.

Iris nodules.

Conjunctival nodules.

Associated Clinical Findings: Cutaneous cafe-au-lait spots.

Fibroma molluscum.

Subcutaneous lipomas.

Sebaceous adenomas.

Facial hemiatrophy.

Growth abnormalities.

Multiple neurofibromas.

Etiology: Familial.

Bibliography

Datelines in Ophthalmology, Vol. 2, No. 1, 1965.

Reese, A.: *Tumors of the Eye*. Paul B. Hoeber, New York, N.Y., 1951.

VOSSIUS' RING

Ocular Findings: Pigmented ring on the anterior lens capsule.

Occurs in young patients after contusions of the globe.

Pigmented ring results partly from the imprint of the pupillary border of the iris and partly from degenerative changes in the lens epithelium.

Tends to disappear with time.

Bibliography

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

VROLIK'S SYNDROME

Similar Condition: *Van der Hoeve Syndrome*.

Ocular Findings: Blue Sclera.

Associated Clinical Findings: Micromelia (apparent smallness of limbs).

Frequent pathologic fractures.

"Rubberhead" (compressible skull).

Usually fatal in youth.

Pathology: A form of osteogenesis imperfecta congenita.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

W**WAARDENBURG SYNDROME**

Synonyms: *Waardenburg-Klein Syndrome, Embryonic Fixation Syndrome*.

Ocular Findings: Hypertelorism with notable lateral displacement of the medial canthi.

Laterally placed lacrimal puncta.

Heterochromia iridis.

Growing together of the eyebrows.

Prematurely gray eyebrows and eyelashes.

Caruncle hypoplasia.

Blepharophimosis may be seen.

Associated Clinical Findings: Congenital deafness.

White forelock.

Brachycaphaly.

Broad, high nasal root.

Protruding lower lip.

Etiology: Familial. Dominant inheritance.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

- Keitel, H.: Peculiar versus diagnostic facies in pediatrics. *Med. Sci.*, 17/6:47-58, 1966.
- Partington, M. W.: Waardenburg's syndrome and heterochromia iridum in a deaf school population. *Excerpta Med.*, 18:530, 1964.
- Waardenburg, P. J.: New syndrome combining developmental anomalies of eyelids, eyebrows, and nose root with pigmentary defects of iris and head hair and with congenital deafness. *Amer. J. Hum. Genet.*, 3:195-253, 1951.

WAGNER'S DISEASE

Synonym: *Hyaloido-retinal Degeneration.*

Ocular Findings: Progressive myopia.

Macular degeneration.

Choroidal sclerosis.

Lattice-like retinal dystrophy with multiple small holes developing in the equatorial and post-ora area.

Retinoschisis.

Scleral ectasia and staphyloma at the posterior pole.

May be associated with cataracta complicata.

Etiology: Autosomal dominant inheritance.

Bibliography

- Alexander, R. L. and Shea, M.: Wagner's disease. *Arch. Ophthalm.*, 74:310-318, 1965.
- Duke-Elder, Sir S.: *Textbook of Ophthalmology*, Vol. 4. C. V. Mosby, St. Louis, Mo., 1949.

WALDENSTRÖM'S SYNDROME

Synonym: *Macroglobulinemia Syndrome.*

Ocular Findings: Hemorrhages and exudates in both retina and choroid.

Occasional papilledema.

Segmentation and clumping of the red blood cells within the conjunctival vessels may be seen.

Associated Clinical Findings: Nasal and oral hemorrhages.

Subarachnoid hemorrhages.

Lymphadenopathy.

Weakness and dyspnea.

Pallor.

Chronic disease mainly affecting males above age fifty.

Pathology: Elevated serum globulin with high sedimentation rate and spontaneous gelification of blood serum at room temperature.

Bibliography

- Ackerman, A.: The ocular manifestations of Waldenström's macroglobulinemia and its treatment. *Arch. Ophthalmol.*, 67:701-707, 1962.
Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
Zlotnick, A.: Macroglobulinemia of Waldenström. *Amer. J. Med.*, 24:461-466, 1958.

WALLENBERG'S SYNDROME

Synonym: *Dorsolateral Medullary Syndrome.*

Ocular Findings: Miosis.

Enophthalmos.

Ptosis.

Diplopia.

Spontaneous homolateral coarse nystagmus.

Associated Clinical Findings: Vertigo.

Dysphagia.

Ipsilateral ataxia.

Muscular hypotonicity.

Ipsilateral loss of pain and temperature sense of the face.

Contralateral partial loss of pain and temperature sense of the trunk and extremities.

Pathologic Etiology: Occlusion of the posterior inferior cerebellar artery.
Prognosis is good.

Bibliography

- Lewis, G. N.; Littman, A.; and Foley, E. F.: Syndrome of thrombosis of posterior inferior cerebellar artery, report of 28 cases. *Ann. Intern. Med.*, 36:592-602, 1952.

WALSH'S RETROBULBAR NEURITIS

Ocular Findings: Axial optic neuritis.

Central visual loss.

Mainly papillomacular bundle involved.

Pathologic Etiology: Usually toxic. Non-infectious.

Bibliography

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

WEBER'S SYNDROME

Synonyms: *Weber-Gubler Syndrome, Cerebellar Peduncle Syndrome.*

Ocular Findings: Ptosis.

Oculomotor paralysis due to homolateral third nerve palsy.

Homolateral fixed, dilated pupil.

Associated Clinical Findings: Contralateral hemiplegia.

Contralateral paralysis of the face and tongue of supranuclear type.

Pathologic Etiology: Lesions of the peduncle, pons, and medulla. Hemorrhage and thrombosis the most frequent cause.

Bibliography

Bedrossian, E. H.: *The Eye*. Charles C Thomas, Springfield, Ill., 1958.

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

Walsh, F. B.: *Clinical Neuro-ophthalmology*, 2nd ed. Williams and Wilkins, Baltimore, Md., 1957.

WEECH'S SYNDROME

Synonym: *Ectodermal Dysplasia Syndrome. See Christ-Siemens-Touraine.*

Ocular Findings: Deficient tearing.

Filiform keratitis.

Corneal dystrophy.

Iris atrophy.

Cataracts have been reported.

Tapetoretinal degeneration.

Loss of brow hair.

Associated Clinical Findings: Hypohydrosis and defective temperature regulation.

Trichosis.

Teeth anomalies (hypodontia).

Dry skin, dry mouth, and atrophic rhinitis.

Patients are "old" looking.
Ears may be pointed.
Low intelligence.
Masculinization may be seen.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148:1-45, Feb., 1966.

WEGENER'S GRANULOMATOSIS

Synonym: *Lethal Midline Granuloma.*

Ocular Findings: Proptosis.

Conjunctivitis.

Scleritis.

Uveitis.

Limbal ulcers.

Orbital cellulitis. Occurs in about fifty per cent of cases due to purulent cellulitis with focal necrosis and erosion into the orbit.

Associated Clinical Findings: Necrotizing granulomatous lesions of the respiratory tract.

Generalized necrotizing vasculitis.

Focal glomerulonephritis.

Pathologic Etiology: Considered to be a hypersensitivity reaction.

Bibliography

Borley, W. E. and Miller, W. W.: Wegener's granulomatosis. Trans. Amer. Acad. Ophthal. Otolaryng., 65:316-323, 1961.

Godman, G. C. and Chung, J.: Wegener's granulomatosis; pathology and review of literature. A.M.A. Arch. Path., 58:533-553, 1954.

McIlvanie, S.: Wegener's granulomatosis. JAMA, 197:90-92, 1966.

Straatsma, B. R.: Ocular manifestations of Wegener's granulomatosis. Amer. J. Ophthal., 44:789-799, 1957.

WEIL'S DISEASE

Synonym: *Epidemic Jaundice.*

Ocular Findings: Intense conjunctival injection.

Subconjunctival capillary hemorrhages.
Jaundice of the bulbar conjunctiva.

Associated Clinical Findings: Jaundice.

Headaches.
Fever.
Hepatomegaly.
Hemorrhagic diathesis.
Abdominal pain.
Muscle weakness.

Etiology: This disease is caused by the leptospira icterohemorrhagiae, a spirochetal disease. Incubation period is five to thirteen days.

Bibliography

Adler, F. H.: *Gifford's Textbook of Ophthalmology*, 6th ed. W. B. Saunders, Philadelphia, Pa., 1959.

WEILL'S SYNDROME

Synonyms: *Weill-Rey Syndrome*, *Adie's Syndrome*, *Pupillotonia*.

Ocular Findings: Pupillary myotonia with slow reaction to light and accommodation.

Clinically appears the same as Adie's Syndrome but is felt to be related to a specific lesion in the infundibular region and the superior peduncle.

Pathologic Etiology: Infundibular region lesions. See Adie's Syndrome.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

WEIL-MARCHESANI SYNDROME

Synonyms: *Marchesani's Syndrome*, "*Inverted Marfan's Syndrome*."

Ocular Findings: Lenticular myopia.
Spherophakia.

Dislocated lenses in many cases.
May have cataracts and glaucoma.

Associated Clinical Findings: Short stature.
Brachydactyly.
Decreased joint flexibility.
Hearing defects.

Pathology: Systemic mesodermal dystrophy. Defective closure of the fetal fissure.

Etiology: Familial.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.
Rosenthal, J. and Kloepper, H. W.: Spherophakia—brachymorphia syndrome. A.M.A. Arch. Ophthalm., 55:28-35, 1956.

WERDNIG-HOFFMANN SYNDROME

Synonyms: *Progressive Myotonic Atrophy, Werdnig-Hoffmann Atrophy.*

Similar Conditions: *Charcot-Marie-Tooth Syndrome, Oppenheim's Disease.*

Ocular Findings: Progressive ocular muscle pareses.
May have nystagmus.
May develop optic atrophy.

Associated Clinical Findings: Onset generally about age six months.
Characterized by progressive generalized weakness and loss of muscle tone in the trunk and extremities.
May have facial and tongue pareses.

Pathology: Lower motor neuron degeneration with diminution in the number of motor cells in the spinal gray matter, with resultant muscular atrophy.

Bibliography

Ford, F. R.: *Diseases of the Nervous System in Infancy, Childhood and Adolescence*, 3rd ed. Charles C Thomas, Springfield, Ill., 1952.

WERNER'S SYNDROME

Synonym: *Progeria*.

Ocular Findings: Bilateral juvenile cataracts of endocrine type.

Occasional bullous keratitis reported.

Trophic defects of the cornea.

Absence of eyelashes and scanty eyebrows.

Associated Clinical Findings: Short stature.

Stretched and atrophic skin.

Ulcerations of the heels, toes and ankles.

Premature baldness.

Hypogonadism.

Thin deformed extremities.

Appears after puberty.

Etiology: Unknown. May be simple recessive as consanguinity of parents is frequent.

Prognosis poor.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

Sorsby, A.: *Modern Ophthalmology*, Vol. 2. Butterworth, Inc., Washington, D.C., 1963.

Thannhauser, S. J.: Werner's syndrome (progeria of adult) and Rothmund's syndrome; two types of closely related heredofamilial atrophic dermatoses with juvenile cataracts and endocrine features; critical study with five new cases. *Ann. Intern. Med.*, 23:559-626, 1945.

WERNICKE'S DISEASE

Synonym: *Nutritional Encephalopathy*.

Ocular Findings: External ophthalmoplegia.

Ptosis.

Paralysis of conjugate gaze.

Horizontal and vertical nystagmus.

Associated Clinical Findings: Mental disturbances.

Ataxia.

Peripheral neuritis.
Most seen in alcoholics.

Pathologic Etiology: Severe thiamine deficiency. Results in destruction of nerve cells and fibers and petechial hemorrhages in the brainstem.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

WERNICKE'S SIGN

Synonyms: *Wernicke's Hemianopic Pupil Sign, Wernicke's Reaction.*

Ocular Findings: In hemianopias, an absent or diminished pupillary reaction when light stimulates the defective side of the retina, and a normal pupillary reaction with light stimulation of the normal side.

The chief usefulness of the Wernicke reaction has been in the differentiation of *tract* hemianopias in which the reaction is positive, from *cerebral* hemianopia in which the pupillary reactions have always been thought to be normal.

It may be demonstrated by alternating a light source from one side to the other and noting the difference in pupillary size.

Bibliography

Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.

WESTPHAL-PILTZ SIGN

Synonym: *Westphal-Piltz Syndrome.*

Ocular Findings: Miosis on forced closure of the lids.

A unilateral constriction of the pupil occurs when the lids are held forcibly apart and an effort is made to close them.

Significance: This is an indication of the integrity of the sphincter pupillae when there is no reaction to light or convergence.

It shows the efferent arc to be intact.

Thought to be due to an overflow of stimuli from the seventh to the third nerve nucleus.

Bibliography

- Cogan, D. G.: *Neurology of the Ocular Muscles*, 2nd ed. Charles C Thomas, Springfield, Ill., 1956.
Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

WEYERS' IRIDODENTAL SYNDROME

Ocular Findings: Iris atrophy.
Glaucoma in severe cases.

Associated Clinical Findings: Ectodermal anomalies.
Oligodontia (few teeth).
Masculinization.

Etiology: Felt to be a hereditary chromosomal abnormality.

Bibliography

- von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

WEYERS-THIER SYNDROME

Synonym: *Oculo-vertebral Dysplasia*.

Ocular Findings: Microphthalmia, usually unilateral.
Unilateral developmental hypoplasia of the orbital bones.

Associated Clinical Findings: Scoliosis and other vertebral anomalies.
Unilateral mandibulo-facial dysostosis.
Macrostomia.

Etiology: Felt to be due to an embryonal oxygen lack.

Bibliography

- von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

WIDMARK'S CONJUNCTIVITIS

Synonym: *Acute Exfoliative Catarrhal Conjunctivitis.*

Ocular Findings: A catarrhal inflammation associated with loss of the epithelium.

In the typical case there is marked hyperemia below the cornea and in the lower tarsal conjunctiva and there are large denuded areas of epithelium which is easily seen with fluorescein staining.

Accompanied by considerable pain.

Etiology: Non specific.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis., Mo., 1965.

WILDER'S SIGN

Ocular Findings: Hesitant jerking or twitching of the globe when moving the eye from extreme abduction to adduction.

Resembles abortive nystagmus.

An inconsistent sign in Graves' disease, but is considered an early sign of exophthalmic goiter.

Associated Clinical Findings: Graves' disease.

Systemic signs of hyperthyroidism.

WILSON'S DISEASE

Synonym: *Hepato-lenticular Degeneration.*

Ocular Findings: Gray-green to golden colored pigmented ring (Kayser-Fleischer) in the periphery of Descemet's membrane.

Occasionally have night blindness.

Associated Clinical Findings: Cirrhosis.

Jaundice.

Dysphagia.

Muscular rigidity.
Drooling.
Coarse tremors.
Ataxia.

Pathology: High copper content in the liver and brain. Low serum uric acid. In the brain, affects mainly the putamen and lenticular nuclei.

Etiology: Familial. Autosomal recessive inheritance.

Bibliography

- Donaldson, D.: Kayser-Fleischer ring in hepatolenticular degeneration (Wilson's Disease). *Arch. Ophthalmol.*, 72:116-117, 1964.
Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.
Hogan, M. J. and Zimmerman, L. E.: *Ophthalmic Pathology*. W. B. Saunders, Philadelphia, Pa., 1962.

WILSON-BROCQ DISEASE

Synonym: *Erythroderma Exfoliativa*.

Similar Condition: *Leiner's Disease*.

Ocular Findings: Exfoliative dermatitis of the lids.

Keratitis and symblepharon may occur.
Corneal sensitivity becomes diminished.
Corneal ulcers may occur.

Associated Clinical Findings: Exfoliative dermatitis usually occurring secondarily to other precedent inflammation.

Characterized by a pink eruption of the skin, followed by scaling and atrophic changes.

Bibliography

- Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 8. C. V. Mosby, St. Louis, Mo., 1965.

WINIWARTER-BUERGER SYNDROME

Synonym: *Thromboangiitis Obliterans*.

Ocular Findings: Retinal vasoconstriction with occasional thrombosis and hemorrhage.

Associated Clinical Findings: Seen in middle-aged patients with thromboangiitis obliterans.

Intermittent claudication.

Cold extremities.

May be complicated by gangrene.

Sensory-motor cerebral defects and psychic symptoms may be seen.

Clinical Note: This condition plays no role in retinal periphlebitis.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche Symptomenkomplexe und Krankheiten. *Klin. Monatsbl. Augenheilk.*, 148:1-45, Feb., 1966.

WYBURN-MASON SYNDROME

Synonym: *Cerebroretinal Arteriovenous Aneurysm Syndrome.*

Similar Condition: *Sturge-Weber Syndrome.*

Ocular Findings: Hemangioma of the disc.

Vision may be impaired.

Homonymous hemianopia.

Ptosis is occasionally found.

Ocular pareses.

Associated Clinical Findings: Midbrain hemangiomas.

Headaches.

Facial palsy.

Speech defects.

Mental retardation is frequent.

Subarachnoid hemorrhages and hydrocephalus may occur.

Bibliography

Duke-Elder, Sir S.: *System of Ophthalmology*, Vol. 3. C. V. Mosby, St. Louis, Mo., 1963.

Wyburn-Mason, R.: On some anomalous forms of anurotic idiocy and their bearing on the relationship of the various types. *Brit. J. Ophthal.*, 27:193-207, 1943.

Z

ZIEVE'S SYNDROME

Synonym: *Hyperlipemia-hemolytic Anemia-icterus Syndrome.*

Ocular Findings: Jaundice frequently appearing initially in the sclera.

Associated Clinical Findings: Usually found in alcoholics.

Epigastric pain.

Jaundice.

Telangiectasia.

Hepatomegaly.

Malaise.

Pathology: Hypercholesterolemia and hemolytic anemia related to later stages of alcoholism.

Bibliography

Durham, R. H.: *Encyclopedia of Medical Syndromes*. Harper and Row, New York, N.Y., 1960.

Zieve, L.: Jaundice, hyperlipemia and hemolytic anemia; a heretofore unrecognized syndrome associated with alcoholic fatty liver and cirrhosis. *Ann. Intern. Med.*, 48:471-496, 1958.

ZINSSER-ENGMAN-COLE SYNDROME

Synonym: *Congenital Dyskeratosis Syndrome.*

Ocular Findings: Bullous conjunctivitis.

Atresia of the lacrimal puncta and resultant epiphora.

Associated Clinical Findings: Skin changes (poikilodermatosis) and nail dystrophy.

Leukoplakia.

Hypogenitalism.

Alterations in the blood picture.

Etiology: Familial. Congenital.

Bibliography

von Grósz, S.: Eponyme-Syndrome. An Autorennamen gebundene augenärztliche

Symptomenkomplexe und Krankheiten. Klin. Monatsbl. Augenheilk., 148-1-45, Feb., 1966.

ZINSSER-THOMSON DISEASE

Synonym: *Infantile Poikiloderma*.

Ocular Findings: Bilateral congenital cataracts.

Associated Clinical Findings: Cutaneous eruptions of the xanthomatous type with zones of atrophy and sclerosis, and zones of depigmentation.

Etiology: Familial. Autosomal recessive.

Bibliography

Francois, J.: *Heredity in Ophthalmology*. C. V. Mosby, St. Louis, Mo., 1961.

SECTION TWO

EPONYMS IN OPHTHALMIC SURGERY

PREFACE TO SECTION TWO

THIS section contains a few selected procedures illustrative of the many eponymic variations in Ophthalmic Surgery and is not intended to be all-inclusive. It is intended only as a guide to some of the more commonly confronted surgical eponyms encountered by the student and resident in eye surgery. For a more detailed and accurate description with relevant illustrations, the reader is referred to the excellent texts and articles referred to in the bibliography.

The procedures described are gathered from many sources, not necessarily the originator's description, and therefore may not agree in all points with what any one particular author describes under the eponym. Also, some of the techniques have been modified by the author. You will note that several of the eponyms described in this section are included primarily for historical reasons. Each reader should therefore be his own editor in the light of his own experience.

The bibliographic references are for further study by the interested reader and are not necessarily the only source for the descriptions given in this section.

SPENCER P. THORNTON, M.D.

A

ATKINSON AKINESIA

Purpose

Akinesia of the orbicularis muscle.

Procedure

1. The point of a No. 22 hypodermic needle, at least 3.5 cm in length, is inserted through the skin and into the orbicularis muscle at the lower edge of the zygomatic bone a little posterior to a vertical line extended from the lateral orbital margin.
2. The shaft of the needle is directed toward the top of the ear, and the anesthetic solution is injected as the needle is directed deeply in the orbicularis near the periosteum.

3. The needle is advanced almost its full length to cover the nerve distribution. Atkinson injects about 3 cc of anesthetic solution, but many ophthalmologists use 10 cc.
4. Firm pressure over the area injected produces a more rapid nerve block.
5. For retrobulbar block, Atkinson recommends a No. 19 needle with a dull point, lessening the possibility of a retrobulbar hemorrhage.

Bibliography

Atkinson, W. S.: *Anesthesia in Ophthalmology*. Publication 251, American Lecture Series, Charles C Thomas, Springfield, Ill., 1955.

B

BARKAN GONIOTOMY

Purpose

To open the filtration angle of the eye with congenital glaucoma.

Indications

Congenital glaucoma from obstruction of persistent embryonic tissue in the angle, and glaucoma associated with the Sturge-Weber Syndrome. This procedure is contraindicated in the late distended stage of buphthalmos in which Schlemm's canal may be obliterated.

Procedure

1. A surgical contact lens is placed over the cornea and physiologic saline is instilled to fill the space between the lens and cornea.
2. The contact lens is held over the nasal side of the cornea to expose the temporal limbus 2 or 3 mm for the introduction of the Barkan goniotomy knife.
3. The cornea is penetrated by the knife somewhere along the temporal limbus and directed across the anterior chamber anterior to the iris.
4. The tip of the blade is inserted for a depth of about $\frac{1}{2}$ to 1 mm in the embryonic tissues to unblock the trabeculum. At least one-third of the circumference should be opened.
5. When the incision is completed, the knife is removed and the anterior chamber is filled with air through the knife incision.

6. More recently, goniotomy knives have been designed which have in their shaft a small metal tube, through which saline may be injected into the anterior chamber as the procedure is performed.

Bibliography

Barkan, O.: Surgery of congenital glaucoma. *Amer. J. Ophthal.*, 36:1523-1534, 1953.
Barkan, O.: Goniotomy. *Trans. Amer. Acad. Ophthal. Otolaryng.*, 59:322-332, 1955.

BEER'S OPERATION

Indication

Iridectomy for restoring visual pathway through updrawn iris.

Result Desired

Surgically-produced pupillary aperture.

Procedure

Described in 1798, Beer incised the limbus with a Beer knife (thin, narrow, straight blade), pulled out a sector of the iris and excised it.

Historical Note

Though designed for the formation of a surgically-produced pupillary aperture, this operation led to the glaucoma iridectomy of Von Graefe in the second half of the nineteenth century.

Bibliography

Sorsby, A.: *Modern Ophthalmology*, Vol. 1. Butterworth, Inc., Washington, D.C., 1963.

BERKE PTOSIS PROCEDURE

Purpose

Resection of the levator by the cutaneous approach.

Indication

Blepharoptosis with a functioning levator muscle.

Procedure

1. A lid spatula is held under the upper eyelid and a skin incision is made 5 mm above the ciliary margin. This is extended through the orbicularis muscle for the length of the lid.

2. The skin and orbicularis are reflected downward from the tarsus almost to the lash line.
3. The eyelid is then pulled downward and the septum orbitale is dissected and cut free from the upper edge of the tarsus.
4. The levator and conjunctiva are buttonholed about 20 mm apart just above the tarsal edge and a ptosis clamp applied.
5. The levator and conjunctiva are cut free from the upper tarsal edge.
6. The conjunctiva is separated from the muscle layer and cut from the ptosis clamp and resutured to the upper tarsal edge.
7. The medial and lateral horns of the levator are cut along with check ligaments.
8. The eyelid is elevated to the desired position at the upper limbus of the cornea. The levator muscle is drawn downward and where it crosses the middle of the tarsus a double-armed suture is directed through the middle third of the levator and tied firmly at this level.
9. Two additional double-armed sutures are similarly inserted through each side of the levator at the same level and tied securely.
10. The muscle is excised below the tied sutures and one needle of each suture is directed through the anterior surface of the tarsus at the level of the lashes, the sutures tied and cut.
11. Three additional double-armed sutures are inserted through the conjunctiva above the tarsus and through the upper tarsal edge and then directed through the levator, and on through the skin edges and tied. The skin edges are united with silk sutures.

Bibliography

- Berke, R. N. and Wadsworth, J. A.: Histology of levator muscle in congenital and acquired ptosis. *A.M.A. Arch. Ophthal.*, 53:413-428, 1955.
- Berke, R. N.: Complications in ptosis surgery. In: Fasanella, R. M., ed., *Management of Complications in Eye Surgery. Avoidance of Pitfalls, etc.* W. B. Saunders, Philadelphia, Pa., pp. 71-101, 1957.
- Berke, R. N.: Results of resection of the levator muscle through a skin incision in congenital ptosis. *A.M.A. Arch. Ophthal.*, 61:177-201, 1959.

BLASCOVICS ECTROPION PROCEDURE

Purpose

Tightening and shortening the lower lid.

Indication

Senile atonic ectropion.

Procedure

1. A skin incision is made about 4 mm below the lower lid margin and extended the length of the lateral half of the lid.
2. The incision is extended upward and outward for about 3 cm.
3. A triangle of skin is excised at the lateral end of the incision with the apex upward. The width of the base determines the amount of slack to be corrected.
4. A full-thickness sector of the lower lid is removed from the lateral half, the width of the resection being determined by the severity of the ectropion.
5. The skin edges are then undermined.
6. The lid marginal defect edges are united with edge-to-edge sutures.
7. The skin flap is slid over to cover the triangular defect laterally and the skin edges united.
8. A pressure dressing is applied and maintained for a day or two.

Bibliography

Blaskovics, L. and Kreiker, A.: *Eingriffe am auge. passim*. Enke, Stuttgart, Germany, 1954.

BUROW'S SKIN TRIANGLES**Purpose**

To facilitate closure of sliding flaps.

Indications

Excess skin and subcutaneous tissue in plastic repair, usually in the angles of pedicle flaps.

Procedure

1. A triangular area of skin on one or both sides of the flap is excised.
2. As the sliding flap is advanced to cover the defect, the Burow's Skin Triangles are closed.
3. The skin edges are united with interrupted sutures.

Bibliography

Burow, A.: Zur blepharoptaslik. Monatschr. med. Augenh. and Chir., 1:57, 1838.

C

CALLAHAN ECTROPION CORRECTION

Purpose

To restore the lid to normal function.

Indication

Senile ectropion with contracting orbicularis.

Procedure

1. A full-thickness wedge is resected from the part of the lid which turns outward the most. The width of the resection is determined by the amount of laxiness of the lid margin.
2. At the edges, the lamellae are halved as in the classic Wheeler procedure.
3. The contracting orbicularis muscle sector is excised on each side of the resection. The orbicularis edges are not sutured.
4. To control the increased horizontal tension upon reunion of the wound edges, a mattress suture is inserted through the tarsus medially and through the orbicularis muscle and skin laterally, and tied over a rubber strip. The margin should be matched perfectly.
5. Interrupted edge-to-edge sutures are used to unite the skin edges.
6. A pressure dressing is applied and maintained for a day or two.

Bibliography

Callahan, A.: Senile ectropion. Analysis of types and surgical correction. *Amer. J. Ophthal.*, 38:787-790, 1954.

D

DAVIEL'S CATARACT OPERATION

Purpose

Restoration of vision by extracapsular cataract extraction. Published 1748.

Indication

Cataract.

Procedure

1. Corneal incision near the lower limbus made by puncture with a sharp, curved knife. This puncture was enlarged to the right

- and left with a blunt curved needle, and the incision to each side was completed with curved convex scissors.
2. A spatula was introduced into the eye to hold the cornea away from the lens. A sharp needle was then used for opening the capsule.
 3. The spatula was next passed between the iris and lens to free any adhesions.
 4. Gentle pressure to dislodge the cataract completed the operation.

Bibliography

Sorsby, A.: *Modern Ophthalmology*, Vol. 1. Butterworth, Inc., Washington, D.C., 1963.

E

ELLIOT CORNEOSCLERAL TREPHINATION

Purpose

Corneosclerectomy for formation of a filtering fistula.

Indication

Chronic open angle glaucoma.

Procedure

1. A large and wide flap of conjunctiva and Tenon's capsule is dissected down to the limbus. The flap should be concentric with the limbus.
2. While the flap is retracted down over the cornea, a corneal splitter is used to split the cornea for about 1 mm from the limbus.
3. A trephine is then applied at the upper limbus to include the posterior part of the split cornea, and the sclera. The trephine is held so that the anterior edge is cut first, insuring a posteriorly hinged disc.
4. As soon as penetration occurs, the iris prolapses into the wound. The disc is then removed with scissors and a peripheral iridectomy performed.
5. The conjunctival flap is replaced and with interrupted sutures, the original incision is united.

Bibliography

Elliot, R. H.: Preliminary note on a new operative procedure for the establishment of a filtering cicatrix in the treatment of glaucoma. *Ophthalmoscope*, 7:804-807, 1909.

ELSCHNIG PROCEDURE (LATERAL CANTHORRHAPHY)**Purpose**

Elevation of the lower lid with permanent adhesions between the upper and lower lids.

Indication

Lids with permanent lagophthalmos. In some instances with marked exophthalmos, and for mild ectropion of the lower lid.

Procedure

1. The upper and lower lids are incised along the intermarginal gray line from the lateral angle toward the nose the length of the desired adhesion.
2. From the posterior lamella of the upper lid, a wedge is excised with the apex placed laterally.
3. A double-armed mattress suture is passed out through the lower lid incision from the conjunctival side of the flap and through a corresponding area of upper lid, orbicularis, and skin, thus filling in the defect in the upper lid. This is tied over a rubber strip on the skin surface.
4. Additional sutures are passed through the lid margins to stabilize the closure.
5. Sutures are removed in five to seven days.

Bibliography

Elschnig, A.: *Augenärztliche Operationslehre*. Springer, Berlin, Germany, 1:309, 1922.

F**FINK SUPERIOR OBLIQUE TUCKING****Purpose**

Shortening of the superior oblique muscle.

Procedure

1. The superior rectus is exposed and dissected free of Tenon's fascia and check ligaments. A muscle hook is then inserted under the superior rectus and it is drawn to the side, exposing the tendon of the superior oblique.

2. The superior oblique is dissected free of fascia and check ligaments for 10 to 15 mm from its insertion.
3. Fink's superior oblique tucker is placed on the superior oblique close to its insertion, and the amount of tucking is determined by the degree of the paralysis, the instrument being adjusted as indicated.
4. Locking sutures are placed through the tendon on each side at appropriate positions. Then a mattress suture is placed through the central portion of the tucked tendon, between the locking sutures.
5. The tucker is removed and additional sutures are placed if necessary. The tucked portion is flattened out to cover the sutures and the superior rectus is allowed to fall back in place over the tuck.
6. It is advisable to abrade the tendon slightly near the sutures to assure good union with the tucked portion.
7. The conjunctiva is united with interrupted edge-to-edge sutures.

Bibliography

Fink, W. H.: *Surgery of the Vertical Muscles of the Eye*. Charles C Thomas, Springfield, Ill., 1962.

FLIERINGA RING TECHNIC

Purpose

Intraocular surgery. To prevent vitreous loss by support of the sclera.

Indications

Any intraocular procedure where vitreous loss may be anticipated.

Procedure

1. The procedure utilizes a stainless steel ring of about 18 mm in diameter (the Flieringa-Bonaccolto ring) which is sutured to the episclera about 3 mm from the limbus encircling the cornea. It is anchored securely in four positions by 4-0 silk suture. (Sharp scleral type needles should be used.)
2. The suture ends are left long so that upward traction during the operation can support the globe firmly and prevent vitreous loss.
3. The ring and sutures are removed after the corneal wound is closed.

Bibliography

Flieringa, H. J.: Procedure to prevent vitreous loss. *Amer. J. Ophthal.*, 36:1618-1619, 1953.

FOX' DOUBLE TRIANGLE PROCEDURE FOR ENTROPION**Indications**

All cases of senile entropion.

Procedure

1. A triangle of skin and muscle lateral to the lateral canthus, measuring 10 to 12 mm across the base (which is up) and about 11 mm vertically is resected.
2. A triangular sector of the tarsoconjunctiva in the center of the lid, base down, 6 to 8 mm at the base is excised. The apex is in the gray line.
3. The edges of both excised triangles are closed and united with interrupted silk sutures, care being taken in the skin closure to secure the muscle fibers as well as the skin.
4. A firm supportive dressing is applied and allowed to remain for forty-eight hours and then changed.

Bibliography

Fox, S. A.: *Ophthalmic Plastic Surgery*, 3rd ed. Grune and Stratton, New York, N.Y., 1963.

FRIEDENWALD-GUYTON SUTURE**Purpose**

Correction of ptosis of the upper lid by attaching it to the frontalis.

Indication

When the levator palpebrae is completely paralyzed.

Procedure

1. Two small stab incisions are made through the skin and muscle of the lid about 5 mm from the margin of the upper lid to the tarsus. The incisions are about 2 cm apart.
2. Two larger stab incisions are made through the skin and muscle to the periosteum just above the eyebrow. These incisions are about 3 cm apart.
3. A large cutting needle threaded with a non-absorbable 2-0 suture

(as cotton or silk) is inserted through the medial brow incision and directed through the frontalis fascia along the periosteum and out through the lateral incision.

4. The long end of the remaining suture is threaded in the cutting needle and passed from the medial brow incision downward through the lid beneath the orbicularis muscle to emerge through the medial lid incision.
5. The needle, rethreaded, is reinserted in the medial lid incision and directed toward the lateral lid incision through which it is drawn out.
6. The needle, rethreaded, is again reinserted and passed upward through the lid to emerge through the lateral brow incision. Now the ends of the sutures are pulled up and after adjusting the proper alignment of the lid margin, the ends are tied deeply in the tissues at the incision.
7. The immediate result should appear as a slight overcorrection.

Bibliography

Friedenwald, J. S. and Guyton, J. S.: Simple ptosis operation; utilization of frontalis by means of single rhomboid-shaped suture. *Amer. J. Ophthal.*, 31:411-414, 1948.

FROST SUTURE

Purpose

Temporary closure of the lower lid.

Indication

To protect the cornea after lid operations, especially for ptosis.

Procedure

1. Two double-armed silk sutures are threaded through rubber strips.
2. The needles are directed through the skin and orbicularis muscle 3 or 4 mm below the margin of the lower lid, then out through the lid margin.
3. The sutures are brought out over the upper lids and directed through the skin and subcutaneous tissues of the eyebrow, and tied over rubber strips.
4. Some ophthalmologists have modified step three, and, holding the suture over the eyebrow, place a piece of adhesive tape over the sutures firmly to hold them without the necessity of directing the needles and sutures through the tissues above.

Bibliography

Frost, A. D.: Supporting suture in ptosis operations. *Amer. J. Ophthalm.*, 17:633, 1934.

G**GRAEFE SECTION****Purpose**

Corneoscleral incision for intraocular surgery.

Procedure

1. The eye is held with fixation forceps applied just below the insertion of the lateral rectus.
2. A Graefe knife is inserted through the conjunctiva just anterior to the lateral rectus insertion and the point introduced through the limbus about 1 mm from the corneoscleral junction.
3. The blade is advanced across the anterior chamber anterior to the iris and the counter-puncture made on the opposite side of the eye so that the blade will come out in the sclera about 1 mm from the corneoscleral junction.
4. With gentle side to side motion the knife is carried upward through the limbus and continued above through the conjunctiva producing a conjunctival flap at the time of the section.

Bibliography

Sorsby, A.: *Modern Ophthalmology*, Vol. 4. Butterworth, Inc., Washington, D.C., 1964.

GUNDERSEN CONJUNCTIVAL FLAP**Indications**

Ulcerative keratopathy with corneal perforations.

Procedure

1. An extremely thin conjunctival flap is prepared in the upper scleral region.
2. A complete peritomy is performed.
3. The involved area of the cornea is excised by a superficial keratectomy. The epithelial tags at the limbus are removed.
4. The flap is drawn downward over the cornea in the prepared area.

5. The lower conjunctival edge is sutured to the cornea with pre-placed silk sutures.
6. The upper edge is sutured to the episclera and Tenons capsule in the area of the peritomy above, leaving the conjunctival blood supply intact at either end.

Bibliography

Gundersen, T.: Conjunctival flaps in the treatment of corneal disease with reference to a new technique of application. *A.M.A. Arch. Ophthal.*, 60:880-888, 1958.

H

HUMMELSCHEIM OPERATION

Purpose

To convert the secondary action of the vertical muscles to abduction by transplantation of their lateral halves to the insertion of the lateral rectus.

Indications

Paralysis of the lateral rectus.

Procedure

1. The lateral rectus muscle is exposed and the amount of resection performed as determined by the degree of deviation; the medial rectus is usually recessed at this time.
2. The resected lateral rectus muscle is laid aside and the insertion of the superior rectus tendon is exposed.
3. The lateral half of the superior rectus tendon is separated from the medial half and detached from the sclera. The strip divided should be about 10 mm long.
4. A locking suture is placed in the free (insertion) end of the muscle strip and passed through the episclera just above the insertion of the lateral rectus and tied.
5. The same procedure is performed on the inferior rectus tendon. The sutured free end of this muscle strip is attached to the episclera at the lower end of the lateral rectus insertion.
6. The resected lateral rectus muscle is resutured to the sclera at the original point of insertion.
7. The medial halves of the superior and inferior rectus muscles are resutured to the temporal end of their insertions.
8. This procedure is usually accompanied by a recession of the medial rectus.

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K

KRONLEIN-BERKE PROCEDURE

Purpose

Resection of the lateral wall of the orbit.

Indications

For removal of orbital lesions which cannot be satisfactorily approached by the anterior or transconjunctival route.

Procedure

1. A lateral canthotomy is made, and the incision is extended directly back toward the ear from the lateral canthal angle. The canthotomy is carried to the periosteum of the lateral orbital rim; the incision to temporalis fascia.
2. The periosteum is incised and the periorbitalis retracted from the lateral orbital wall, toward the apex of the orbit.
3. The bone incisions are outlined by incising the periosteum of the outer surface of the lateral orbital wall. The upper line should begin at the orbital rim 10 mm above the lateral orbital tubercle, and the lower one just above the temporal process of the zygomatic bone.
4. The bone is cut with a Stryker saw so that a wedge-shaped section of bone extending about 12 mm from the orbital rim can be broken out and reflected, being attached by periosteum.
5. If more exposure is needed posteriorly, more of the lateral wall can be removed with rongeur forceps.
6. Closure is effected by holding the bone in place and suturing the edges of periosteum. Then the extensions of the lateral canthal ligament are reattached to the periosteum along the orbital rim. The overlying tissues are closed in layers.

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- Kronlein, R. V.: Zur pathologie and operative behandlung der dermoidcysten der orbita. Beitr. klin. Chir., 4:149-163, 1888-89.

KUHNT-SZYMANOWSKI PROCEDURE**Purpose**

To shorten and strengthen the support of a ptosed lower lid.

Indications

Senile atonic ectropion.

Procedure

1. The lower lid is split into two laminae by an incision through the marginal gray line and extended well below the lower tarsal border. The incision is begun lateral to the punctum and extended for about 20 mm, past the lateral canthal angle with a skin incision that follows the normal lower lid curve.
2. From the lateral end of the skin incision a downward vertical incision of about 15 mm is made. The skin flap thus created is undermined in continuity with the outer lamina of the lid.
3. A triangle of tarsoconjunctiva is excised from the area where the ectropion is most pronounced, with the base of the triangle at the lid margin. The excised triangle should be of such size that when the edges are approximated, the lid will lie firmly against the globe.
4. The edges of the tarsoconjunctival lamina are then united with mattress sutures, usually of gut sutures with the knots placed on the anterior surface of the tarsus.
5. The skin flap is then pulled laterally and that portion which extends beyond the vertical incision is marked off and removed. The ciliated lid margin that now extends beyond the lateral canthal angle is excised.
6. A holding suture is placed at the apex of the lateral edge of the skin flap and the vertical incision is closed. Then the skin edges and lid margin are closed with interrupted silk sutures.

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L

LAGLEYZE ENTROPION PROCEDURE

Purpose

Correction of cicatricial entropion and trichiasis.

Procedure

1. The upper lid is everted on a lid retractor and held in this position by a suture through the lid margin.
2. Five special half-curved "ski" needles are threaded onto a 50 cm length of non-absorbable suture allowing equal lengths of suture between needles.
3. The conjunctiva and tarsus of the upper lid are incised about 3 mm from the margin for almost the length of the lid. A small strip of conjunctiva and tarsus is excised, the length of the tarsus.
4. The middle needle is inserted through the conjunctival surface just above the upper tarsal edge. The needle passes through the conjunctiva, Mullers muscle and the levator and along the anterior surface of the tarsus to the incision; the needle is then directed through the mid-thickness of the tarsus below the incision and out through the ciliary margin.
5. The remaining needles are spaced evenly and passed similarly through the tissues to make their exit through the ciliary margin. When all of the suture has been drawn out, the needles are then cut off. The paired suture ends are tied over a piece of rubber tubing or gauze to maintain eversion of the lid.
6. These sutures are allowed to remain in place for four to five weeks.

Bibliography

Lagleyze: Operation for entropion and trichiasis by a new method. *Ann. Ophthal.*, 14:473-477, 1905.

LAGRANGE-FORONI OPERATION

Purpose

To produce a filtering cystoid cicatrix and full iridectomy.

Indication

Glaucoma, open angle type.

Procedure

1. The iris is contracted preoperatively with eserine sulfate $\frac{1}{4}$ per cent.
2. A thick, wide limbus based conjunctival flap is made.
3. The incision into the anterior chamber is made through the posterior part of the scleral trabeculae in the limbal tissues.
4. A scalpel is used to excise a crescent of sclerocorneal tissue from the anterior lip of the incision at the base of the conjunctival flap.
5. As the conjunctival flap is held downward, the iris is grasped at the sphincter, and pulled out so that when the full sector iridectomy is performed, it will match the position of the sclerectomy. After first cutting the iris to make an iridodialysis, it is torn from its base and the second cut is made.
6. The iris pillars are then swept out of the incision and the conjunctival flap then closed.

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M

McREYNOLDS PTERYGIUM TRANSPLANTATION**Purpose**

Transplantation and emplacement of the head of the pterygium in a pocket of conjunctiva.

Procedure

1. The head of the pterygium is dissected off the cornea by a superficial keratectomy beginning in clear cornea just beyond the apex of the pterygium.
2. A conjunctival incision is made along the lower border of the pterygium from the corneoscleral junction outward. The length of this incision should equal the width of the pterygium.
3. A pocket is prepared in the subconjunctival area below the incision of sufficient depth to accommodate the pterygium.
4. Both needles of a double-armed suture are passed through the apex of the pterygium several millimeters apart and the suture

ends are drawn through to let the loop remain against the apex.

5. The needles are passed into the subconjunctival pocket and out through the conjunctiva and the pterygium is pulled down into the pocket. The suture is then tied to hold the head of the pterygium in place.

Bibliography

McReynolds, J. O.: Pterygium operation. *Southern Med. J.*, 28:823-828, 1935.

MOTAIS PTOSIS CORRECTION

Purpose

To attach the superior rectus muscle to the tarsal plate of the upper lid.

Procedure (modified)

1. An incision is made in the line of the normal lid fold for about 25 mm.
2. The lower flap, including skin and orbicularis, is dissected down to the lash line.
3. The upper flap is freed to the upper tarsal edge, and the conjunctiva at the edge is opened with a 10 mm incision.
4. The globe is rotated downward and the conjunctiva opened over the insertion of the superior rectus. This muscle is exposed for 10 mm.
5. Three double-armed sutures are inserted through the muscle 3 mm from the insertion and tied. The muscle is then cut from the globe leaving a stump through which are passed two double-armed sutures which are passed through the muscle about 5 mm behind the three previous sutures and tied.
6. The conjunctival wound is closed around the tongues of muscle which now protrude about 5 mm.
7. The superior rectus is pulled through the conjunctival lid incision and the needles passed over the tarsus through the lid margin exiting through the gray line where they are tied.
8. Mattress sutures of silk are passed through the lid skin incision to anchor the superior rectus to the skin at the skin fold.

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Motais, M.: Opération du ptosis pour la greffe tarsienne d'une languette du tendon du muscle droit supérieur. Bull. et Mem. Soc. Franc. d'ophth., 15:208-217, 1897.

N

NAFFZIGER TRANSCRANIAL ORBITAL DECOMPRESSION

Indications

Cranial dysostosis, some types of dysthyroid exophthalmos.

Procedure

1. Coronal incision and anterior cranial approach (transcranial but extradural).
2. The orbit is unroofed, permitting the orbital contents to proptose into the anterior cranial space, effecting orbital decompression.

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O

O'BRIEN AKINESIA

Purpose

To obtain akinesia of the orbicularis.

Procedure

1. The needle is introduced 1 cm anterior to the tragus of the ear, directly over the condyloid process of the mandible. The needle is passed directly inward toward the bone, injecting about 2 cc of anesthetic solution into the deep tissues in the plane of the nerves.
2. The needle is withdrawn after injection of only 2 cc. If the needle has been introduced properly and the tip in the region of the periosteum as the anesthetic is injected, lid paralysis should appear within one minute.
3. If paralysis is not complete because of inadequate placement, it is better to proceed immediately to the Van Lint method rather than to repeat the original injection.

Bibliography

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O'CONNOR CINCH OPERATION

Purpose

Shortening of an extraocular muscle.

Procedure

1. The extraocular muscle is exposed and then stretched between two strabismus hooks.
2. The muscle is divided into four equal bundles and then, beginning with the upper bundle, several strands of dermal suture are wound around that bundle three or four times (enclosing 3 or 4 mm of muscle) away from the insertion.
3. The dermal suture is then passed across the second muscle section and wound around it an equal number of times back toward the insertion.
4. The sutures are then passed across the third muscle bundle and again wound outward three or four loops, then passed over the bottom (fourth) muscle section and wound back toward the insertion.
5. The dermal strands are then pulled taut, producing a muscle roll around the dermal strands. The upper end of the dermal suture is cut 5 or 6 mm above the edge of the muscle.
6. The lower ends of the dermal sutures are brought out through the wound and tucked into the inferior cul-de-sac and the conjunctiva is closed.
7. The dermal sutures are pulled out in two weeks.

Bibliography

O'Connor, R.: Cinch shortening loop in surgery of extraocular muscles. *Western J. Surg.*, 39:670-680, 1931.

S

SNELLEN ENTROPION SUTURE

Purpose

To produce eversion of the lower lid.

Indication

Spastic entropion.

Procedure

1. Two or three double-armed 5-0 sutures are used as mattress

sutures by placing the needles through the conjunctiva in the lower cul-de-sac 3 or 4 mm apart.

2. The needles are then passed subcutaneously upward to emerge through the skin just below the lid border.
3. Each set of sutures is tied over a rubber strip.
4. The sutures are removed in two weeks.

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V

VAN LINT AKINESIA

Purpose

To obtain akinesia of the orbicularis.

Procedure

1. A hypodermic needle, No. 22, 3.5 cm long, is inserted about 1 cm lateral to the orbital margin in line with the lower orbital rim and after injecting a small amount of anesthetic subcutaneously, the needle is directed into the deep tissues just below the orbital margin near the bone, and advanced along the lower orbital margin to the medial third.
2. About 2 cc of anesthetic solution is injected as the needle is withdrawn and as the needle approaches its point of insertion it is not withdrawn but again advanced into the deep tissues along the lateral rim toward the eyebrow and another 2 cc of anesthetic injected along this tract.
3. It may be necessary to inject more anesthetic a little lower in the cheek for complete akinesia.
4. If the injections are placed close enough to the orbital margin so as to catch the emerging sensory nerves, anesthesia as well as akinesia of the lids is produced. However, swelling around the orbit may be a problem, and Van Lint's method requires more anesthetic than those placed more proximally with attendant risk of toxicity.

Bibliography

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VERHOEFF CATARACT EXTRACTION**Purpose**

Sliding technique of lens delivery.

Procedure

1. After making a limbal section to open the anterior chamber, a full sector iridectomy is performed at the twelve o'clock position.
2. The Verhoeff capsule forceps are used to grasp the lens equator at the site of the iridectomy.
3. Lifting traction is exerted above with the forceps and external pressure upward is applied to the cornea below. As the lens is delivered, the forceps are gently moved from side to side.

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Verhoeff, F. H.: New operation for removing cataracts with their capsules. *Trans. Amer. Ophthal. Soc.*, 25:54-64, 1927.

W**WHARTON JONES V-Y PLASTY****Purpose**

Correction of cicatricial ectropion by a sliding flap.

Indication

Mild central ectropion.

Procedure

1. A V-shaped skin incision is made from the eyelid margin converging to an apex below the involved area. The width of the ciliary margin base is determined by the extent of scarring.
2. The triangle of skin is undermined from the apex to the lid margin to completely expose the scarred subcutaneous tissues.
3. All possible scar tissue is excised.
4. The edges of the wound are undermined on each side to form freely movable flaps.
5. With relaxation of the lid, the V-flap retracts. The movable flaps are brought together at the base and sutured to fill in the gap to the apex of the now retracted V-flap.
6. Skin edges are now approximated, resulting in a "Y" closure.
7. In recent years, this procedure has been used much less often.

Bibliography

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WHEELER DISCISSION**Purpose**

To open an optical area through a membranous or secondary cataract.

Procedure

1. With a Wheeler discission knife, an entrance is made into the cornea 1 mm in from the limbus at twelve o'clock.
2. The knife point is carried across the anterior chamber and beneath the inferior pupillary border of the iris.
3. The membrane is incised from behind the iris inferiorly upward through the pupil by using the corneal wound of entrance as a fulcrum and bringing the knife handle up as it is withdrawn so that the blade makes an incision through both the membrane and the anterior vitreous in an arc, emerging at the upper part of the membrane. Wheeler specified that this should be done with speed.
4. The knife is withdrawn as the incision is completed.

Bibliography

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WHEELER HALVING PROCEDURE**Purpose**

Repair of lacerated eyelid without lid notching.

Procedure

1. Beginning at the edge of the lid laceration a skin incision is made about 2 mm below the ciliary margin and extended 3 to 5 mm away from the wound through skin and orbicularis.
2. A skin incision is made from the lateral end of the infraciliary incision to the tip of the lid laceration creating a triangle of skin and muscle. This anterior lamina is removed.
3. An equal amount of the conjunctiva and tarsus are removed from the opposite side, including a strip of the ciliary margin

matching the length of the preserved portion opposite so that when brought together will form an overlapping joint.

4. A double-armed mattress suture is passed through the posterior conjunctival and tarsal layer so as to come out in the center of the excised triangle and then is continued through the opposite side of the wound through the muscle and skin so that the lid edges and skin margins match perfectly.
5. The sutures are snugged up and tied over a rubber strip.
6. The skin edges are then closed by interrupted silk sutures.

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Callahan, A.: *Surgery of the Eye: Diseases*. Charles C Thomas, Springfield, Ill., 1956.
Wheeler, J. M.: *Collected Papers*. Columbia University Press, New York, N.Y., 1939.

WHEELER'S TARSORRHAPHY (LATERAL ANGLE CANTHORRHAPHY)

Purpose

To produce intermarginal lid adhesions for permanent closure of the eyelids in cases of permanent lagophthalmos, mild ectropion of the lower lid, and in some cases of exophthalmos.

Procedure

1. An incision is made along the upper and lower lids in the marginal gray lines to a depth of 2 or 3 mm.
2. Beginning on the conjunctival surface of the upper lid, double-armed sutures are directed into the upper lid incision, then directed through the posterior lid flap and then out of the skin of the upper lid. This pulls the posterior flap from the lower lid into the pocket created in the upper lid.
3. Sutures are tied over a rubber strip and a mild pressure dressing applied.
4. Sutures are removed in five to seven days.

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WIES ENTROPION REPAIR

Purpose

Eversion of the eyelid margin.

Indication

Senile cicatricial entropion and other types of entropion.

Procedure

1. An incision is made completely through the lower lid 3 mm below the lid margin and extended the entire length of the lid.
2. Three double-armed mattress sutures are inserted through the lower conjunctival edge and tarsus, continued through the incision and directed to include the skin and subcutaneous tissue above the incision anteriorly.
3. The sutures are drawn snugly and tied, thereby attaching the palpebral conjunctiva and tarsus to the eyelid skin and everting the lid margin.
4. The skin incision is closed with additional silk sutures.

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Z

ZIEGLER CAUTERY PUNCTURE**Purpose**

To turn the lid margin toward its normal position by contracting upon cicatrication after the burn.

Indication

In ectropion, to turn the lid margin in; in entropion, to turn the lid margin outward.

Procedure for Ectropion

1. Anesthesia is induced by topical instillations and injection of local anesthetic.
2. The lid is everted, and at about 5 mm from the margin a pointed electrocautery, heated to high degree is pressed into the conjunctiva and through the tarsus.
3. The cautery is not allowed to reach the skin.
4. Enough punctures are made to counteract the deformity. Sometimes this requires many applications.

5. An antibiotic ointment is instilled and a moderate pressure dressing is applied for twenty-four hours.

Procedure for Entropion

1. Lid is anesthetized as described above.
2. With a through-and-through mattress suture or a lid clamp, the lower lid is put on a stretch. A pointed electrocautery, heated to high degree is pressed into the skin and subcutaneous tissues about 5 mm from the margin. The tarsus is not cauterized.
3. The number of punctures needed varies as described above.
4. An antibiotic ointment is instilled and a moderate pressure dressing is applied for twenty-four hours.

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